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Total *En Bloc* Spondylectomy of C3: A New Surgical Technique and Literature Review

Totální „en bloc“ spondylektomie C3: nová chirurgická technika a přehled literatury

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ABSTRACT

PURPOSE OF THE STUDY

Radical resection of a vertebra is reserved only for specific tumors that invade the surrounding tissues and recur when not removed completely. The vertebra may be removed using a piecemeal technique or *en bloc*, using only two (in thoracolumbar spine) or more osteotomies (in cervical spine). We present our technique of *en bloc* resection of subaxial cervical vertebra for Ewing's sarcoma of C3, with preservation of all nerve roots and both vertebral arteries. To our knowledge, this surgical technique has not been reported in the English literature. The aim of this study is to describe the new technique of radical resection of subaxial cervical vertebra.

MATERIAL AND METHODS

A transoral biopsy of tumor tissue anterior to C2-C3 was performed in 8-year old boy, revealing a diagnosis of Ewing's sarcoma. The patient was started on neoadjuvant chemotherapy. After 6 chemotherapy cycles with the VIDE regimen, the soft-tissue component completely regressed, with the only a residual deposit in C3 vertebral body. Based on further multidisciplinary meeting, an *en bloc* spondylectomy of C3 was recommended, preferably with preservation of nerve roots and vertebral arteries. In August 2014, prior to the planned surgery, we performed another thorough examination of the patient using plain films, CT and MRI. Neither angiography nor embolization was performed.

DESCRIPTION OF SURGICAL TECHNIQUE

The first stage of the operation consisted of resection of the posterior structures. We exposed the posterior elements of C2 to C4 by the mid-line incision. The C3 arch was without pathological changes. After partial resection of the C2 inferior and C4 superior articular processes we performed bilateral osteotomy in the region of the pedicle adjacent to the arch with a chisel and removed the whole of the C3 posterior arch. Subsequently we perforated the transverse foramina close to the pedicle, using fine Kerrison rongeurs. The lateral parts around vertebral arteries were left *in situ.* In the next step we used instrumentation with polyaxial screws to stabilize the C2-C4 section.

After 19 days we performed the second stage surgery from an anterior approach with the removal of the anterior and lateral parts of the vertebra. We made a transverse incision anterior to the sternocleidomastoid between the internal carotid artery and the trachea on the right side at the level of C3 to expose the spine. We resected C2-C3 and C3-C4 intervertebral discs and then performed osteotomy with fine Kerrison rongeurs on both sides, again, close to the vertebral body. Subsequently, the vertebral body was released and extracted *en bloc*. In the next step, both vertebral arteries were mobilized and shifted medially and the lateral portions of the transverse processes were released and removed *en bloc.* The empty space was filled with solid allograft and the C2-C4 levels were bridged by the cervical plate in 2+1+2 configuration.

RESULTS

There were no complications during both surgeries. The follow-up CT examination 4 months after the operation revealed a clear bone fusion of C2-C4, both anteriorly between vertebral bodies and posteriorly between the arches. Clinically the patient has reached 8 month follow up and had no complaints, both he and his parents were satisfied. Physiotherapy is proceeding according to plan. The patient remains under supervision at our centre.

DISCUSSION

Total *en bloc* resection of a subaxial cervical vertebra with preservation of neural and vascular structures has been described in the English literature only once. In 2007 was published a total *en bloc* resection of C5 for chordoma, preserving the above mentioned structures. Authors removed the lamina *en bloc* after bilateral osteotomy. Transverse foramina were perforated by the Gigli saw and removed in piecemeal fashion, including the posterior tubercle. In the next step, they removed the vertebral body and the anterior tubercle from the anterior approach. However, their treatment differs from the technique described here and does not correspond fully to the principle of *en bloc* resection. Our surgical technique is based on a similar principle of performing several osteotomies without the use of high speed burr, while preserving all neural and vascular structures. The difference can be particularly seen in the approach to remove lateral parts of the transverse foramen, which are surrounding the vertebral arteries. We consider it as ideal to split the cervical vertebra by smooth cuts into four parts and remove them *en bloc*.

CONCLUSION

Total *en bloc* spondylectomy of a subaxial cervical vertebra with preservation of vertebral arteries and nerve roots is a radical surgery that should be used to treat only the most serious conditions. The risk of neurological deficit is outweighed by the benefits of oncological radicality. This new surgical technique has not yet been described and it is clear, that a larger cohort of patients is necessary to assess and potentially modify this technique so that it can be used more frequently in the future.

Key words: *en bloc* spondylectomy, total spondylectomy, cervical spine, vertebrectomy.

INTRODUCTION

Radical resection of a vertebra is reserved only for specific tumors that invade the surrounding tissues and recur when not removed completely. The vertebra may be removed using a piecemeal technique or *en bloc*, using only two (in thoracolumbar spine) or more osteotomies (in cervical spine). Techniques of complete *en bloc* removal of a vertebra from a combined or a single posterior approach in the region of thoracolumbar spine have been repeatedly described in the literature (1, 3, 11, 17). However, in the cervical spine, only a few case studies of partial *en bloc* resection (5, 10, 12) and one case of total *en bloc* resection of the subaxial cervical vertebra (8) were reported. *En bloc* resection in the subaxial cervical region may be performed with sacrificing of the unilateral vertebral artery and nerve roots or with their preservation. We present our technique of *en bloc* resection of subaxial cervical vertebra for Ewing's sarcoma of C3, with preservation of all nerve roots and both vertebral arteries. To our knowledge, this surgical technique has not been reported in the English literature.

A CASE STUDY AND DESCRIPTION OF SURGICAL TECHNIQUE

An 8-year old boy was repeatedly examined in the local hospital for pain and stiffness of the cervical spine in autumn 2013. In the beginning of 2014, the pain was getting more intense and from March it also persisted during the night. The patient was subsequently examined by the general orthopaedic surgeon. Radiographs showed increased sclerosis of C3 and MRI examination of the cervical spine was recommended, which was performed in April 2014. It revealed an expansive process between the C2-C4 vertebral bodies, surrounding the vertebral artery on the left, extending as far as the spinal canal and compressing the spinal cord at the C2-C4 level; the spinal cord showed no signs of myelopathy (Fig. 1). Due to suspected malignancy, the patient was referred to Pediatric Hematology and Oncology department of our hospital and at the multidisciplinary meeting a biopsy was suggested. A transoral biopsy of tumor tissue anterior to C2-C3 was performed in April, revealing a diagnosis of Ewing's sarcoma. The tumor was classified IIB according to the Enneking staging system (9) and the modified Weinstein Boriani Biagini (4) staging involved zones 1 to 10, layers A to D and F, levels C2 to C4. The patient was started on neoadjuvant chemotherapy. After 6 chemotherapy cycles with the VIDE regimen, the soft-tissue component completely regressed, with the only a residual deposit in C3 vertebral body. Based on further multidisciplinary meeting, an *en bloc* spondylectomy of C3 was recommended, preferably with preservation of nerve roots and vertebral arteries. In August 2014, prior to the planned surgery, we performed another thorough examination of the patient using plain films, CT and MRI (Fig. 2). Neither angiography nor embolization was performed. After chemotherapy the modified Weinstein Boriani Biagini (4) staging was: zones 5 to 8, layers B and C, level C3 (4).

OPERATIVE TECHNIQUE

The patient under general endotracheal anesthesia was placed prone on a standard operating table with his head fixed by an adhesive plaster in neutral position of the cervical spine. The first stage of the operation consisted of resection of the posterior structures. We exposed the posterior elements of C2 to C4 by the midline incision. The C3 arch was without pathological changes. After the partial resection of the C2 inferior and C4 superior articular processes we performed bilateral osteotomy in the region of the pedicle adjacent to the arch using a flexible thin chisel with the bevel edge turned ventrally (Synthes – chisel handle with changeable blades) in the direction from top and from outside, and we removed the whole C3 posterior arch in one piece. Subsequently we perforated the transverse foramina close to the pedicle, using the fine Kerrison rongeurs. The lateral parts around

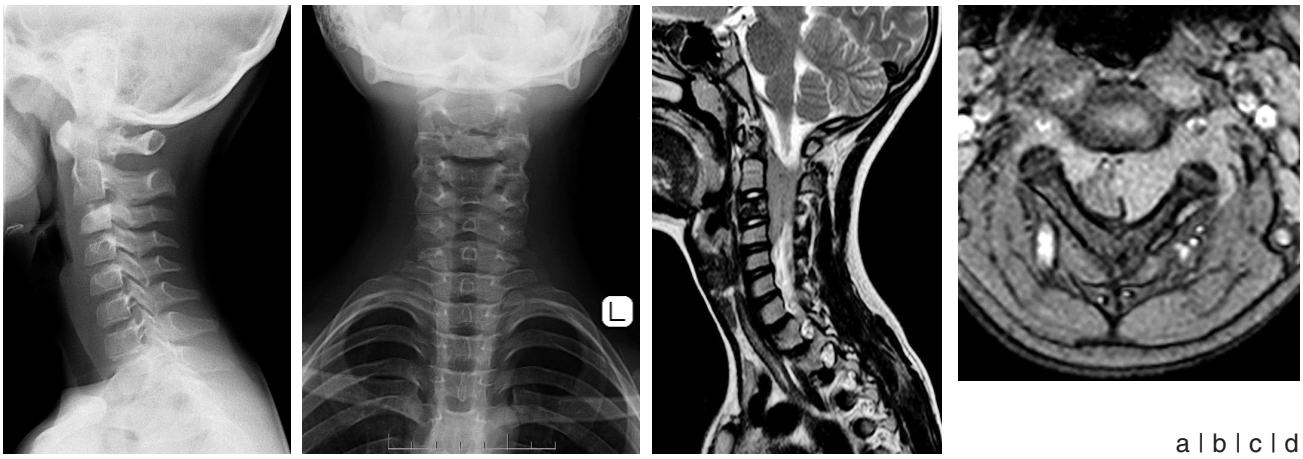

Fig. 1. An 8-year old boy with Ewing's sarcoma of C3 before neoadjuvant chemotherapy: a – lateral plain radiograph of the cervical spine, b – anteroposterior plain radiograph of the cervical spine, c – sagittal T2-weighted MRI scan of the cervical spine, d – axial T2-weighted MRI scan at C3 level.

Fig. 2. Findings after neoadjuvant chemotherapy: a – lateral plain radiograph of the cervical spine, b – anteroposterior plain radiograph of the cervical spine, c – sagittal T2-weighted MRI scan of the cervical spine, d – axial T2-weighted MRI scan at C3 level, e – sagittal CT reconstruction of the cervical spine, f – coronal CT reconstruction of the cervical spine, g – axial CT section at C3 level.

vertebral arteries were left *in situ* (Fig. 3). A specimen of the posterior arch was sent for histological examination. In the next step we used S4 Cervical fixation system with polyaxial screws (Aesculap, Germany) to stabilize the C2-C4 section. Transpedicular 4mm screws were inserted into the C2 and lateral mass 4mm screws into the C4 utilizing Judet and Magerl techniques respectively. A crosslink was used to connect both rods to enhance stability. Posterior fusion of C2-C4 was performed using allograft bone from the bone bank and deep drain was inserted before closure of the wound. The operative time was 195 minutes and the blood loss 2000 ml. The patient spent 4 days on the pediatric ICU.

After 19 days we performed the second stage surgery from an anterior approach with the removal of the anterior and lateral parts of the vertebra. The patient under general endotracheal anesthesia was placed supine on a standard operating table and with support of the cervical spine, the head was fixed by adhesive plaster in a slight extension. We made a transverse incision anterior to the sternocleidomastoid between the internal carotid artery and the trachea on the right side at the level of C3 to expose the spine (standard Smith-Robinson approach). No macroscopic changes were manifested on the C3 vertebral body but only coarse fibrous tissue surrounding the vertebra. We resected C2-C3 and C3-C4 intervertebral discs and then performed osteotomy with fine Kerrison rongeurs on both sides, again, close to the vertebral body. Subsequently, the vertebral body was released and extracted *en bloc*. In the next step, both vertebral arteries were mobilized and shifted medially and the lateral portions of the transverse processes were released and removed *en bloc* (Fig. 4). To achieve superior oncological radicality is necessary to remove all four parts in one piece. The empty space was filled with solid allograft and the C2-C4 levels were bridged by the Trinica plate (Zimmer, USA) in 2+1+2 configuration (Fig. 5). Finally a suction drain was inserted and the wound closed in layers. The operative time of the anterior surgery was 245 minutes and the blood loss 700 ml. After the surgery, the patient was again transferred to ICU and returned back to the ward on 2nd postoperative day. Histological examination of the C3 vertebra was without any tumor cells. Hyperplastic bone beams and haematopoiesis were found in the bone marrow and some of the intramedullary spaces were filled with fibrous tissue. No associated complications were recorded during the two operations or in the early postoperative period.

POSTOPERATIVE CARE AND FOLLOW UP

The patient was wearing a custom-made cervical brace for 3 months and underwent 10 chemotherapy cycles with the VAC regimen in Pediatric Hematology and On-

cology department. Follow-up X-rays taken at 3, 6 and 12 weeks showed unchanged position of the fixation elements. The follow-up CT examination 4 months after the operation revealed a clear bone fusion of C2-C4, both anteriorly between vertebral bodies and posteriorly between the arches (Fig. 6). Clinically the patient has reached 8 month follow up and had no complaints, both he and his parents were satisfied. Physiotherapy is proceeding according to plan. The patient remains under supervision at our centre.

DISCUSSION

Radical resection of a vertebra, in part or as whole of one or more vertebrae is an extensive operation. It is well documented and discussed particularly in the region of the thoracolumbar spine. Radicality of *en bloc* resection of the affected region significantly increases the patient's chances for a long-time survival (1, 3, 11, 17). Depending on the type and extent of the tumor pathology, the choice is made of a technique preserving nerve roots or sacrificing them for the sake of oncological radicality. In the region of the atlantoaxial complex and subaxial cervical spine, these surgical procedures are quite rare and were described only in a few case reports (2, 5, 6, 12). Anatomical relations are more intricate here mainly due to the vertebral arteries passing in the osseous canal. Sacrificing or preserving of the non-dominant vertebral artery is another issue in the decision-making algorithm. The choice of appropriate surgical technique is influenced also by the extent of pathology, particularly by the presence of an extraosseous component and its relation to the vertebral arteries and nerve roots.

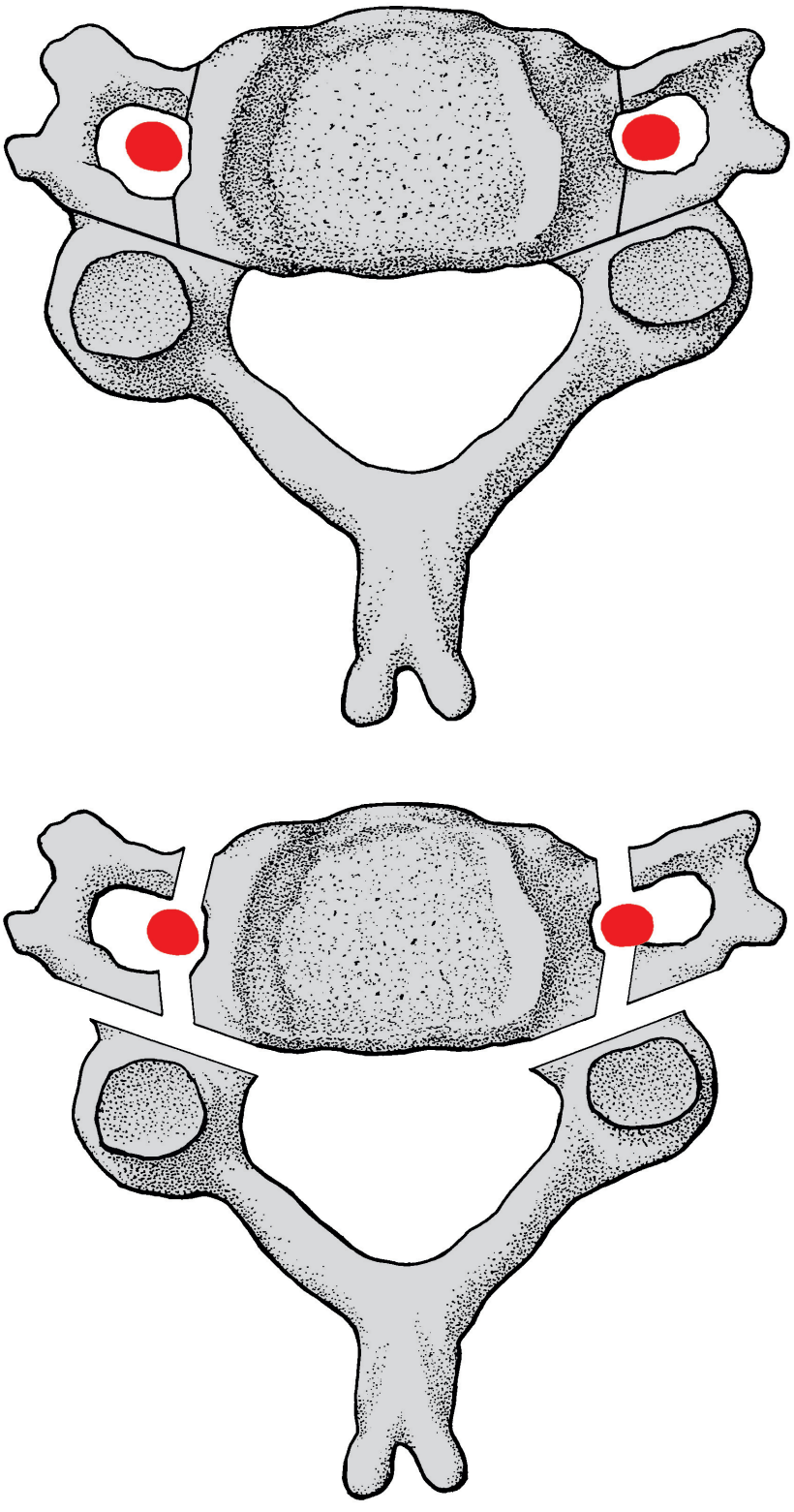

Fig. 3. Illustrations of the osteotomies before and after division.

Complete resection of a cervical vertebra in a piecemeal fashion is the most frequently used technique and is widely documented in the literature (7, 13–16, 18). The most common diagnosis is chordoma with other diagnoses being less frequent. Intralesional resection is associated with high risk of local recurrence. Wang et al. (18) reported 5 patients with total intralesional resection of a cervical vertebra for chordoma. Simsek et al. (13) described two cases of a total intralesional spondylectomy, consisting in one case in extraction of C3 for chondrosarcoma and of C4 for metastasis of the lung adenocarcinoma in the other. Cohen et al. (7) published a multi-level total spondylectomy of C5-C7 for osteosarcoma, with piecemeal extraction of the posterior portion, *en bloc* removal of the anterior portion and again piecemeal removal of the lateral one. Partial *en bloc* resection technique is especially suitable in man-

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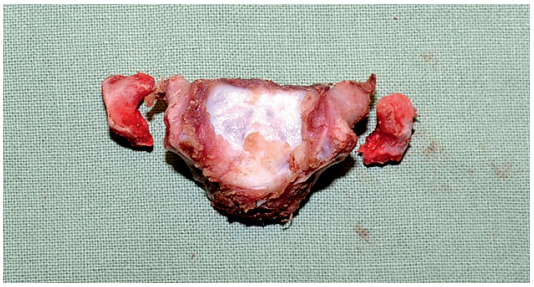

Fig. 4. Intraoperative findings: a – lateral plain radiograph, b – antero-posterior plain radiograph, c – plain radiograph of the removed specimen, d – macroscopic appearance of the removed specimen.

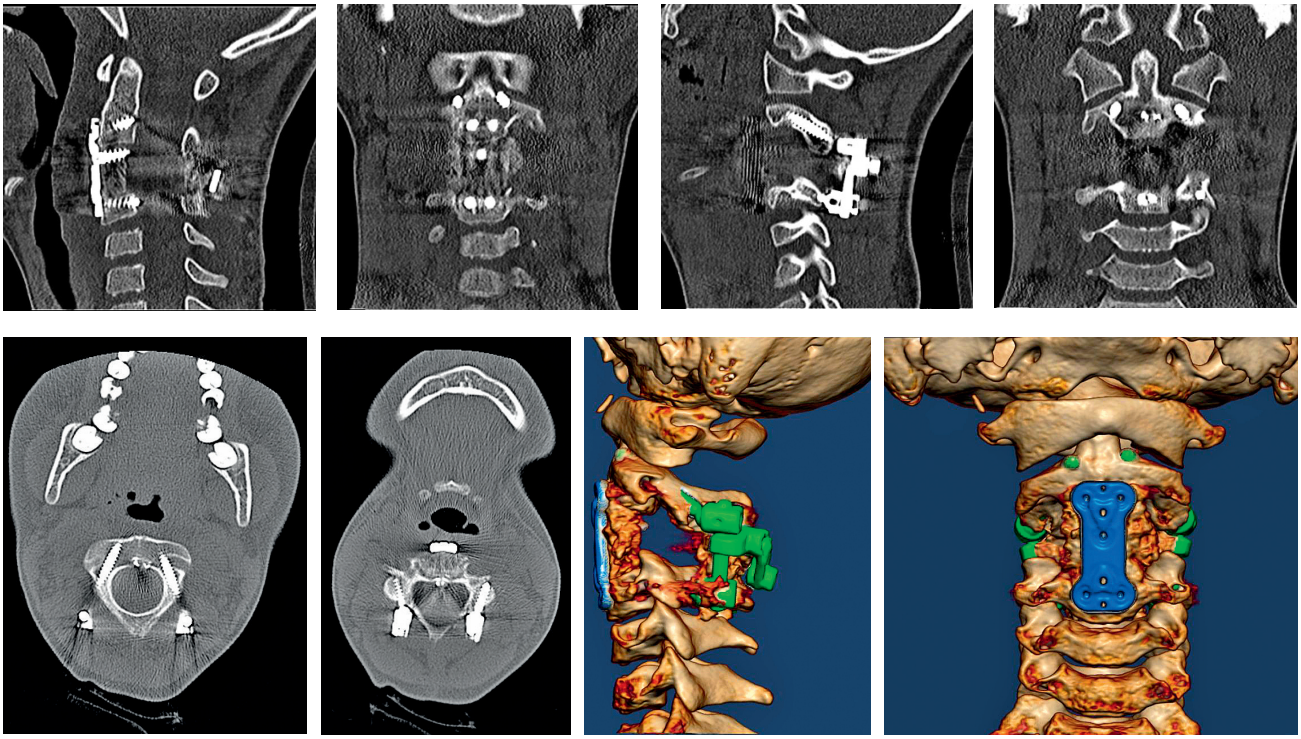

Fig. 5. CT examination immediate post-surgery: a – sagittal CT reconstruction of the midline, b – coronal CT reconstruction at the graft level, c – parasagittal CT reconstruction, d – coronal CT reconstruction behind the graft, e – axial CT section at C2 level, f – axial CT section at C4 level, g – 3D CT reconstruction - lateral view, h – 3D CT reconstruction - anterior view.

agement of tumors with the extraosseous component or of multi-level lesions. In patients in whom ligation of one vertebral artery is considered, it is recommended to perform occlusion test to reduce the risk of brain damage. Rhines et al. (12) in their well-known study reported *en bloc* spondylectomy of C2-C4 for chordoma, combined with ligation of one vertebral artery and dividing of roots on one side. Chou et al. (5) published a multi-level *en bloc* resection of cervical vertebrae in three patients for chordoma. They also resected nerve roots and one vertebral artery. Another description of a similar surgical procedure for chordoma was presented by Cloyd et al.

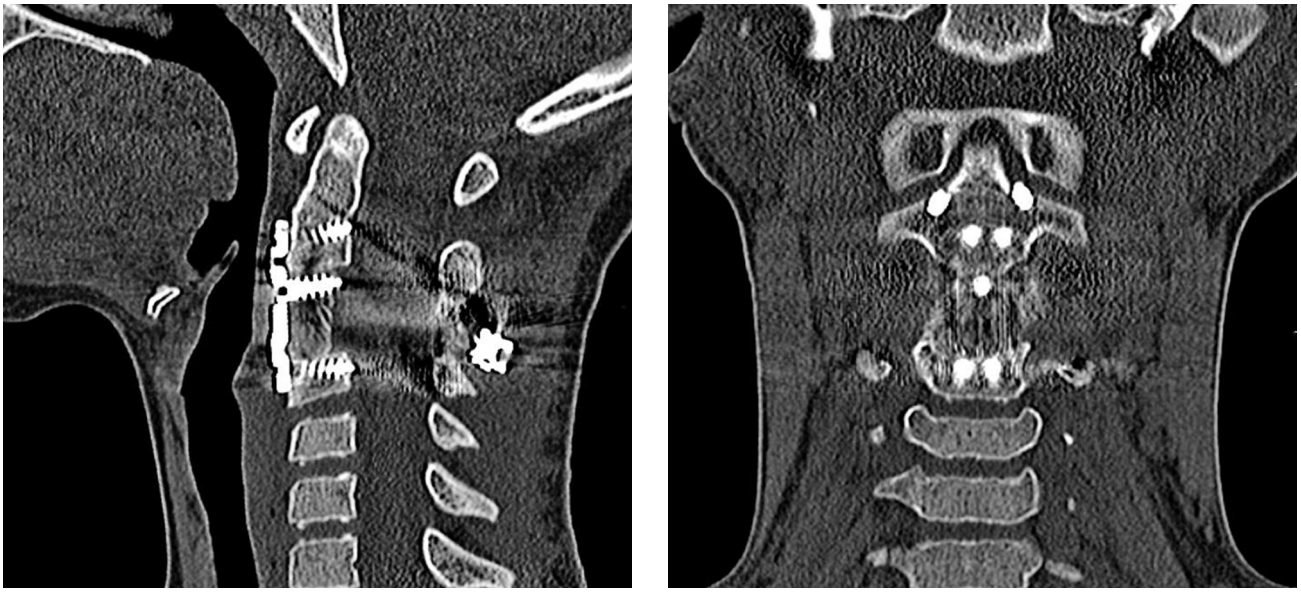

Fig. 6. Follow-up CT scan after 4 months of surgery with clear anterior and posterior bony fusion: a – sagittal CT reconstruction of the cervical spine, b – coronal CT reconstruction of the cervical spine.

(6), who in one patient performed a multi-level *en bloc* resection of C3-C6, again with ligation of nerve roots and vertebral artery. These cases, however, did not accomplish a total removal of the vertebra as whole compartment. Guppy et al. (10) reported a total *en bloc* resection of C2-C3, with sacrificing of nerve roots and vertebral artery on one side, and removal of the bone surrounding the vertebral artery by ultrasonic aspirator on the other. For laminectomy and for removal of the compartment bony structures remote from the tumour they used a high-speed burr. We do not use high speed burr in malignant tumors because we feel it contradicts the principle of oncological radicality.

Total *en bloc* resection of a subaxial cervical vertebra with preservation of neural and vascular structures has been described in the English literature only once. In 2007, Currier et al. (8) published a total *en bloc* resection of C5 for chordoma, preserving the above mentioned structures. They removed the lamina *en bloc* after bilateral osteotomy. Transverse foramina were perforated by the Gigli saw and removed in piecemeal fashion, including the posterior tubercle. In the next step, they removed the vertebral body and the anterior tubercle from the anterior approach. This type of surgery requires tumours confined to intraosseous space and compartments. However, their treatment differs from the technique described here and does not correspond fully to the principle of *en bloc* resection. Our surgical technique is based on a similar principle of performing several osteotomies without the use of high speed burr, while preserving all neural and vascular structures. The difference can be particularly seen in the approach to remove lateral parts of the transverse foramen, which are surrounding the vertebral arteries. We consider it as ideal to split the cervical vertebra by smooth cuts into four parts and remove them *en bloc*.

Removal of a vertebra results in a severe spinal instability, requiring replacement of the missing bony and ligamentous structures. Some authors prefer to perform anterior surgery first (14). However, most of them choose posterior resection and stabilization as the primary procedure in order to ensure a higher degree of stability (5, 6, 8, 10, 15, 16). Posterior fixation is usually performed with polyaxial screw-rod systems (5, 6, 12, 14-16). The extent of fixation is governed by the scope of resection, type of tumor, bone quality and, last but not least, by the established practice of the given centre. Occipitocervical or occipitocervicothoracic fixation is chosen in case of multi-level resections, poor bone quality and in chordomas with high risk of local recurrence (6, 10, 12, 14). Short segment fixation (one level above and one level below) is suitable in case of one-level resection and good bone quality ensuring proper purchase of fixation elements (8, 14, 15). The anterior vertebral column is in most cases replaced by mesh or expandable titanium cages bridged by a plate (5, 6, 10, 12, 14-16), although some authors use tricortical bone graft harvested from the pelvis (8). In pediatric patients, we prefer a structural allograft, bridged and fixed by a plate.

CONCLUSION

Total *en bloc* spondylectomy of a subaxial cervical vertebra with preservation of vertebral arteries and nerve roots is a radical surgery that should be used to treat only the most serious conditions. The risk of neurological deficit is outweighed by the benefits of oncological radicality. Due to the complexity of such procedure, it is best addressed in specialized centers where adequate multidisciplinary cooperation is available. This new surgical technique has not yet been described and it is clear, that a larger cohort of patients is necessary to assess and potentially modify this technique so that it can be used more frequently in the future.

Abbreviations:
VIDE – vincristine, ifosfamide, doxorubicin, etoposide,
VAC – vincristine, actinomycin D, cyclophosphamide,
ICU – intensive care unit.

References

1. ABE, E., SATO, K., TAZAWA, H., MURAI, H., OKADA, K., SHIMADA, Y., MORITA, H.: Total spondylectomy for primary tumor of the thoracolumbar spine. Spinal Cord, 38: 146–152, 2000.
2. BAILEY, Ch. S., FISHER, Ch., G., BOYD, M. C., DVORAK, M. F. S.: En bloc marginal excision of a multilevel cervical chordoma. J. Neurosurg. Spine, 4: 409–414, 2006.
3. BORIANI, S., BANDIERA, S., BIAGINI, R., BACCHINI, P., BORIANI, L., CAPPUCCIO, M., CHEVALLEY, F., GASBARRINI, A., PICCI, P., WEINSTEIN, J. N.: Chordoma of the mobile spine: Fifty years of experience. Spine, 4: 493–503, 2006.
4. CHAN, P., BORIANI, S., FOURNEY, D. R., BIAGINI, R., DEKUTOSKI, M. B., FEHLINGS, M. G., RYKEN, T. C., GOKASLAN, Z. L., VRIONIS, F. D., HARROP, J. S., SCHMIDT, M. H., VIALLE, L. R., GERSZTEN, P. C., RHINES, L. D., ONDRA, S. L., PRATT, S. R., FISHER, C. G.: An assessment of the reliability of the Enneking and Weinstein-Boriani-Biagini classifications for staging of primary spinal tumors by the Spine Oncology Study Group. Spine (Phila Pa 1976), Feb 15; 34: 384–391, 2009.
5. CHOU, D., ACOSTA, F. Jr., CLOYD, J. M., AMES, C. P.: Parasagital osteotomy for en bloc resection of multilevel cervical chordomas. J. Neurosurg. Spine, 10: 397–403, 2009.
6. CLOYD, J. M., CHOU, D., DEVIREN, V., AMES, C. P.: En bloc resection of primary tumors of the cervical spine: report of two cases an systematic review of the literature. Spine J., 9: 928–935, 2009.
7. COHEN, Z. R., FOURNEY, D. R., MARCO, R. A., RHINES, L. D., GOKASLAN, Z. L.: Total cervical spondylectomy for primary osteogenic sarcoma. J. Neurosurg. Spine, 97: 386–392, 2002.
8. CURRIER, B. L., PAPAGELOPOULOS, P. J., KRAUSS, W. E., UNNI, K., K., YASZEMSKI, M. J.: Total en bloc spondylectomy of C5 vertebra for chordoma. Spine, 32: E294–E299, 2007.
9. ENNEKING, W. F.: A system of staging musculoskeletal neoplasms. Clin. Orthop. Relat. Res., 204: 9–24. 1986.
10. GUPPY, K. H., CHAKRABARTI, I., ISAACS, R. S., JUN, J. H.: En bloc resection of multilevel high-cervical chordoma involving C-2: new operative modalities. J. Neurosurg. Spine, 19: 232–242, 2013.
11. MARMOR, E., RHINES, L. D., WEINBERG, J. S., GOKASLAN, Z. L.: Total en bloc lumbar spondylectomy. J. Neurosurg. Spine, 95: 264–269, 2001.
12. RHINES, L. D., FOURNEY, D. R., SIADATI, A., SUK, I., GOKASLAN, Z. L.: En bloc resection of multilevel cervical chordoma with C2 involvement. J. Neurosurg. Spine, 2: 199–205, 2005.
13. SIMSEK, S., BELEN, D., YIGITKANLI, K., ACIDUMAN, A., BAVBEK, M.: Circumferential total resection of cervical tumors: report of two consecutive cases and technical note. Turkish Neurosurgery, 19: 153–158, 2009.
14. SUCHOMEL, P., BUCHWALD, P., BARSA, P., FROEHLICH, R., CHOUTKA, O., KREJZAR, Z., SOURKOVÁ, P., ENDRYCH, L., DZAN, L.: Single-stage total C-2 intralesional spondylectomy for chordoma with three-column reconstruction. J. Neurosurg. Spine, 6: 611–618, 2007
15. ŠTULÍK, J., KOZÁK, J., ŠEBESTA, P., VYSKOČIL, T., KRYL, J., PELICHOVSKÁ, M.: Total spondylectomy of C2: a new surgical technique. Acta Chir. orthop. Traum. čech., 74: 79–90, 2007.
16. ŠTULÍK, J., KOZÁK, J., ŠEBESTA, P., VYSKOČIL, T., KRYL, J., KLÉZL, Z.: Total spondylectomy of C2: report of three cases and review of the literature. J. Spinal Disord. Tech., 23: E53–E58, 2010.
17. TOMITA, K., KAWAHARA, N., BABA, H., TSUCHIYA, H., FUJITA, T., TORIBATAKE, Y.: Total en bloc spondylectomy: A new surgical technique for primary malignant vertebral tumors. Spine, 3: 324–333, 1997.
18. WANG, Y., XIAO, J., WU, Z., HUANG, Q., HUANG, W., ZHU, Q., LIN, Z., WANG, L.: Primary chordomas of the cervical spine: a consecutive series of 14 surgically managed cases. J. Neurosurg. Spine, 17: 292–299, 2012.

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