Clinical case

Spontaneous oral extrusion of an acrylic vertebral reconstruction 12 years after a vertebrectomy for an Ewing's sarcoma of the cervical spine: A case report

Expulsion orale spontanée d'une reconstruction acrylique vertébrale 12 ans après une vertérectomie pour un sarcome d'Ewing cervical : cas clinique

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A B S T R A C T

Introduction. – Primary Ewing sarcoma of the cervical spine is rare, particularly in children population. The surgical management remains a challenge to associate the best oncological resection and to prevent spinal deformity. The situation is complicated owing to paucity of adapted instrumentation and their possible interactions with the growing bone.

Clinical presentation. – We described the case of a young 19-year-old woman admitted for an oral extrusion of a bone polymethyl methacrylate (PMMA) allograft 12 years after a C4 circumferential vertebrectomy for primary Ewing’s sarcoma. The vertebral anterior reconstruction was slowly repulsed by the growing spine giving way to an autologous bone without kyphosis deformation.

Conclusion. – Bone reconstruction remains a challenge after extensive oncological resection particularly in cervical spine of children. Anterior and posterior instrumentation must be associated. The growing spine is not a good host for PMMA allograft and autograft seems to be preferred for anterior column fusion. In spite of the good oncological results, the authors raise the long-term issue of PMMA for vertebral reconstruction in young patients. With a long follow-up, they showed that posterior rigid fixation might prevent the cervical kyphosis.

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R É S U M É

Introduction. – Le sarcome d’Ewing du rachis cervical est particulièrement rare chez l’enfant. Sa prise en charge chirurgicale reste complexe afin d’associer une résection oncologique tout en prévenant l’apparition d’une déformation rachidienne. La situation est compliquée par le manque de matériel adapté et leur possible interaction avec l’os en croissance.

Présentation clinique. – Nous rapportons le cas d’une jeune femme de 19 ans qui a été admise pour l’expulsion par la bouche d’une allogreffe en polymethyl methacrylate (PMMA) 12 ans après une vertérectomie circonférentielle de C4 pour un sarcome d’Ewing. La reconstruction antérieure a été lentement repoussée par le rachis en croissance laissant place à un os autologue sans déformation en cyphose.

Conclusion. – La reconstruction osseuse reste un véritable challenge après des résections larges à visée carcinologique, particulièrement pour le rachis cervical de l’enfant. Une instrumentation antérieure et postérieure doit être associée. Le rachis en croissance n’est pas un hôte favorable pour recevoir une allogreffe de PMMA. L’autogreffe doit être privilégiée pour la fusion de la colonne antérieure. Malgré un bon résultat carcinologique, les auteurs soulèvent le problème du devenir à long terme du PMMA pour la reconstruction vertébrale chez les patients jeunes. Ils montrent également avec un suivi important que la fixation postérieure prévient la déformation en cyphose.

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1. Introduction

The Ewing’s sarcoma family of tumours (ESFT) was first described by James Ewing in 1921 [1]. ESFT represents 3% of all
the paediatric malignant tumours and is the most common primary malignant paediatric spine tumour with an incidence of 3.5% to 10% of all cases [2].

The most common signs are pain followed by neurological deficits. About a quarter of patients display metastases at the time of diagnosis: lungs (50%), bone (25%) and bone marrow (20%).

Treatment is based on combined neoadjuvant chemotherapy, radiotherapy and wide surgical excision. However, rapidly progressing neurological disorders constitute an indication for immediate surgery [3]. Surgical resection with adequate margins improves survival among patients with osteosarcoma, but increases the risk of spinal instability and requires stabilization.

We report an exceptional paediatric case of cement dislodgment and its oral reject 12 years after a C4 vertebrectomy for a metastatic ESFT.

This case highlights the problem of the interaction between polymethyl methacrylate (PMMA) and the growing spine.

2. Case report

2.1. History and examination

This 7-year-old girl presented with a one-week history of a right arm weakness. She complained of neck pain evolving since 3 months. At the admission the examination revealed a torticollis with a right biceps weakness. An MRI study showed a large enhanced epidural tumour extending from C3 to the C4-C5 intervertebral disc space. Severe spinal cord compression was showed behind the body of C4 (Fig. 1). As she presented with an acute neurological deficit, she underwent a surgery few days after her admission: C4 corporectomy and reconstruction with PMMA. Simultaneously, a C2 to C6 posterior instrumentation was performed to reduce the risk of kyphosis deformation. Immediate postoperative lateral X-rays were satisfying: the anterior reconstruction of C4 reinforced by two Kirschner’s pins and the posterior instrumentation were well positioned (Fig. 2). The pathological evaluation confirmed the diagnosis of Ewing’s sarcoma with positive immunoreactivity for anti-MiC2. Bone marrow and lung locations were found during staging. Radiotherapy was performed and the patient was started on cycles of cyclophosphamid, adriamycine, vepeside and isofosfamid. The treatment was completed by autologous stem cells transplantation. The postoperative course was uneventful. A regular clinical and radiological follow-up was arranged. Eleven years after surgery, while she was asymptomatic, a discrete anterior migration of the reconstruction was misdiagnosed on the lateral X-Ray (Fig. 3). Twelve years after surgery, she was admitted for a spontaneous expulsion of a foreign body during a meal. No warning symptoms as dysphagia, dysphonia, dyspnea, cough or cervical pain were reported. During neurological examination, no sign of spinal irritation or radicular disorder were found. A slight wound of the posterior wall of the hypopharynx was
diagnosed at nasofibroscopy and spontaneous healing was observed. The anterior migration evolved to the complete expulsion of PMMA secondary to a spontaneous ossification of the removal cavity. This aspect of “new vertebra” provided an anterior arthrodesis from C3 to C5, completing the spinal stabilization by the posterior instrumentation (not shown). Sixteen years after the vertebral the surgery and 4 years after the expulsion a new X-ray was realized showing the same aspect (Fig. 4). The patient was cured and no spine kyphosis was observed. The posteriors columns were fused.

Macroscopic examination of the foreign body confirmed that it was the acrylic reconstruction (Fig. 5).

3. Discussion

Primary osteosarcomas of the cervical spine in child have been described in few cases. Radicular pain is one of the most common symptoms at presentation (85%) and neurological deficit is present in 40% of the cases [3]. Current treatment consists in neoadjuvant chemotherapy administered after biopsy but before surgery to decrease the surgical morbidity by reducing the tumor volume [4]. When the patient presents with an acute neurological disorder, a surgical decompression is preferred to a close biopsy [3]. In our case, a surgical decompression associated to a combined anterior-posterior approach was decided. This approach facilitates the radical resection and the circumferential stabilization reduces late kyphotic deformity [5]. After an anterior vertebrectomy, several reconstruction techniques are feasible: bone autograft, intervertebral spacer or acrylic cement [6]. Some authors have described PMMA vertebral reconstruction for spinal metastasis in adult people with short life expectancy [7]. PMMA increases the surrounding bone turnover [8]. It activates peripheral blood monocytes promoting a periosteosis inflammation and a bone resorption by osteoclast activation. This aseptic loss of the surrounding bone had probably disconnected the graft of the bone and contributed to its gradual rejection by the growing spine. In our case, the vertebral reconstruction was completely excluded by a slow fistulation to the pharynx and was orally rejected despite its size and the two Kirchner’s pins anchored in C3 and C5. The graft dislodgment is a well-know complication of the cervical reconstruction especially

Fig. 3. 11 years postoperative lateral X-ray: anterior migration of the acrylic vertebral replacement and exclusion of the C3 pin. Note the posterior fusion between C3 and C5.

Radiographie cervicale de profil 11 ans après la chirurgie montrant le déplacement antérieur de la vertèbre acrylique et l'exclusion de la broche de Kirschner de C3. Fusion des corps vertébraux de C3 et C5.

Fig. 4. 16 years postoperative and 4 years after vertebral reconstruction expulsion lateral X-ray: good spine stability. Absence of kyphosis. Radiographie cervicale de profil 16 ans après la chirurgie et quatre ans après l'expulsion de la reconstruction vertébrale: bonne stabilité rachidienne. Absence de cyphose.

Fig. 5. Lateral view of the acrylic vertebral replacement with the two Kirchner’s pins after it oral expulsion. Vue latérale de la vertèbre acrylique avec les deux broches de Kirschner après l'expulsion orale.
in case of multilevel vertebrectomies without internal stabilization [9]. In a serie, Zileli et al. [10] have described one case of an ante-
dorial cervical plate (with its screws) extruded after a spondylectomy for a cervical chordoma.

This case highlights the difficulty for pediatric cervical recon-
struction and stabilization. After an anterior corpectomy, the
another column needs to be reconstructed. The PMMA vertebral
reconstruction was performed because of the lack of anterior cervi-
cal instrumentation available at the time of diagnosis (1990s). Even
though fixation implants were available at the thoracic and lumbar
pediatric segments [11], no implants were specially designed for
the cervical segment. Some studies have described cervical fusion
with tricortical bone graft and anterior miniplate and screws after
a cervical corpectomy in child harboring an eosinophilic gran-
uloma [12]. The authors describe a satisfactory vertebral fusion
with autogenous iliac crest and we should have preferred a bone
autograft to PMMA.

Bone autograft reconstruction in primary Ewing sarcoma of the
cervical spine after a C5 resection has been previously published
[13] with a good oncological result. In a more recent review the
authors considered the bone autograft, particularly the vascular-
ized graft, as an optimum solution for the surgical reconstruction
in children with osteosarcoma [14].

However, despite a properly bone fusion, the results have
been presented respectively at only 18 and 24 months after the
reconstruction. A longer follow-up is necessary to measure spine
deformity. In fact, an extensive vertebral resection could lead to
instability and a posterior instrumentation is recommended to pre-
vent kyphotic deformity. After 16 years of follow-up we did not
observe cervical kyphosis despite the expulsion of the vertebral
reconstruction. Our case is a good illustration that posterior inter-
nal fixation is a good strategy to prevent kyphosis in the growing
spine. Posterior cervical instrumentation with lateral mass screw
or hooks and rods could be used. The vertebral fusion and the stabili-
ization may prevent deformation but could affect future growth of
the cervical spine for some authors. Another group has described
the use of absorbable anterior cervical plate for cervical anterior
fusion [15] without alignment and fusions complications. These
plates were developed to avoid an irregular growth of the spine
that could be feared with titanium plates.

These promising plates could be used when a circumferential
fixation is needed.

4. Conclusion

The treatment of ESFT of the growing spine is challenging for surgical excision and vertebral reconstruction and stabilization
steps. En bloc total vertebrectomy could represent an option
in some well-defined cases. Wide excision is fundamental for
the prognosis but it should not compromise the neurological
function and stability of the spine. Extensive resections could be
performed, involving complex spinal reconstructions to prevent
kyphotic deformation. This case illustrates the need for a preop-
erative discussion on equipment to use depending on patient’s age
and disease prognosis, particularly in children.

Depending on the situation, the surgeon has the choice to use
different devices such as autografts, prostheses or composites.
Despite this good oncological result and the absence of kypho-
sis deformity, PMMA is not adapted for the growing spine. Today
a bone autograft and absorbable plate should be associated to a
cervical posterior fixation.

Disclosure of interest

The authors declare that they have no conflicts of interest concern-
ning this article.

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