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

R. Faguer*, D. Petit, P. Menei, H.-D. Fournier

Department of neurosurgery, University hospital, 4, rue Larrey, 49933 Angers cedex 9, France

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ABSTRACT

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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RÉSUMÉ

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 a 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 ifosfamid. 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

Fig. 1. Sagittal T1-weighted with gadolinium MRI of the cervical spine showing an anterior epidural soft tissue tumour compressing the spinal cord from C3 to C5. Note also the pre-s spinal soft tissue infiltration.

IRM sagittale T1 avec injection de gadolinium montrant une compression médullaire antérieure étendue de C3 à C5 avec envahissement des tissus pré-vertébraux.

Fig. 2. Two days after surgery, plain lateral X-ray showing the acrylic vertebral replacement reinforced by two Kirschner’s pins and the posterior instrumentation (Cotrel-Dubousset’s technique).

Radiographie cervicale postopératoire de profil montrant la reconstruction vertébrale acrylique ancée par deux broches de Kirschner et l’ostéosynthèse postérieure de type Cotrel-Dubousset.
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 posterior 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 periprosthesis 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 fistulization 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
in case of multilevel vertebrectomies without internal stabilization [9]. In a serie, Zileli and al. [10] have described one case of an anterior cervical plate (with its screws) extruded after a spondylectomy for a cervical chordoma.

This case highlights the difficulty for pediatric cervical reconstruction and stabilization. After an anterior corpectomy, the anterior column needs to be reconstructed. The PMMA vertebral reconstruction was performed because of the lack of anterior cervical instrumentalisation 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 fixation with tricortical bone graft and anterior miniplate and screws after a cervical corpectomy in child harboring an eosinophilic granuloma [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 vascularized 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 prevent 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 internal 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 stabilization 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 preoperative 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 kyphosis 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 concerning this article.

References