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CASE REPORT

A novel fixation technique using anterior C1 screw in a pediatric solitary cervical spinal juvenile xanthogranuloma

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Abstract

Purpose Juvenile xanthogranuloma (JXG) presenting as solitary vertebral body lesion is infrequently seen and usually limited to one or two levels. We report a case of an isolated JXG with extensive cervical spinal (bony and extradural) involvement in a 6-year-old child. There was a diagnostic dilemma as the radiologic and intraoperative picture resembled tuberculosis. The spinal reconstruction was also challenging due to involvement of multiple vertebral levels and necessitated an anterior C1 screw.

Methods The lytic lesion was multicompartmental, involving the craniovertebral junction and the subaxial spine (till C6 vertebral body) and extending into the retropharyngeal space. Noticeably, an associated thoracic syringomyelia was also present. Near-total excision of the lesion and 360° spinal fixation was performed using fibular strut graft. The graft was cranially anchored to the C1 anterior arch, thereby sharing the load with the posterior occipito-cervical instrumentation in order to avoid a construct failure due to cantilever effect.

Results At 12-month follow-up, the patient had good clinico-radiologic outcome with evidence of bony fusion and resolution of syrinx.

Conclusion The report highlights the diagnostic dilemma of JXG lesion on both the radiology and surgery and discusses the challenges in the management and the relevant literature. The described technique can be a viable option in pediatric tumors with extensive C2 vertebral body involvement. Occasionally, extradural compression can have associated syrinx formation and the primary treatment per se could tackle the underlying syringomyelia.

Keywords Juvenile xanthogranuloma · Non-Langerhans cell histiocytosis · Craniovertebral junction · Cervical spine · Fibular graft · Vertebral body · C1 screw

Introduction

Juvenile xanthogranuloma (JXG) is a rare, benign, non-Langerhans cell histiocytic proliferative disorder and represents 0.5% of pediatric tumors [1]. It commonly presents as a self-limiting, isolated skin lesion. Occasionally, the disorder can affect subcutaneous tissue, eye, viscera, central nervous system and bones. Such an extracutaneous involvement is seen in approximately 5–10% of cases [2–4].

Spine is an unusual site for JXG. Very few reports describe JXG which presented as isolated vertebral body lesions, mostly confined to one or two vertebral bodies [3–6]. We hereby report a case of pediatric solitary JXG with extensive involvement of the cervical spine along with spread into the retropharyngeal space. The relevant differential diagnosis and the management issues have been discussed.

Case report

A 6-year-old boy who is a diagnosed case of Klippel–Feil syndrome presented with 2-year history of neck tilt and recent-onset dysphagia. He also had complaint of mild neck pain. General physical examination showed pectus excavatum. His neurology was normal. Computed tomography

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(CT) of the cervical spine showed an osteolytic destruction involving the C2, fused C3–C5 and C6 vertebral bodies (Fig. 1). The C1 arch and uppermost portion of dens were intact. Magnetic resonance imaging (MRI) demonstrated a contrast-enhancing extradural soft tissue lesion intending the cord and bulging out anteriorly into the retropharyngeal space. An associated thoracic syringomyelia was noted.

A CT-guided biopsy of the lesion was inconclusive. In view of the spinal compression and indefinite biopsy report, an excision of the lesion was planned (by senior author PS). The child underwent excision of the mass through an anterior approach along with posterior stabilization that included the occiput, C1 lateral mass, C7 and T1 pedicle screws. During surgery, it was seen that the prevertebral fascia was plastered. On incising it, a dirty-white, flaky material was noted within the lesion. The mass was near-completely excised leaving the portion adjacent to the vertebral arteries. An autologous fibular graft was then fashioned to bridge the bony defect and fixed with cortico-cancellous screws, the cranial one extending from the graft into the C1 anterior arch, and the caudal screw into the C7 body.

Intraoperative frozen sections showed histiocytic collection and occasional multinucleated giant cells. Considering the high endemicity of tuberculosis in our area, the child was discharged on antituberculous therapy. The final histopathological examination of the paraffin-embedded sections, however, turned out to be JXG (Fig. 2).

The child was on Philadelphia collar for 3 months. At 12-month follow-up, he was doing well without any evidence of recurrence. Repeat imaging showed resolution of the syrinx and good bony fusion (Fig. 1).

Discussion

According to the recent classification, the histiocytic disorders consist of five groups such as Langerhans-related (L group), cutaneous and mucocutaneous (C group), malignant histiocytoses (M group), Rosai–Dorfman disease (R group), and hemophagocytic lymphohistiocytosis and macrophage activation syndrome (H group) [7]. JXG is the most common form of cutaneous histiocytosis (C group). The extracutaneous variety is biologically different from cutaneous JXG and is classified in L group [7]. Histologically, JXG is composed of characteristic Touton giant cells in a background of mononuclear cells which are seen in about 85% of cases [2]. These giant cells may be less pronounced in extracutaneous lesions [2]. It is important to distinguish JXG from Langerhans cell histiocytosis, as the latter shows more aggressive clinical

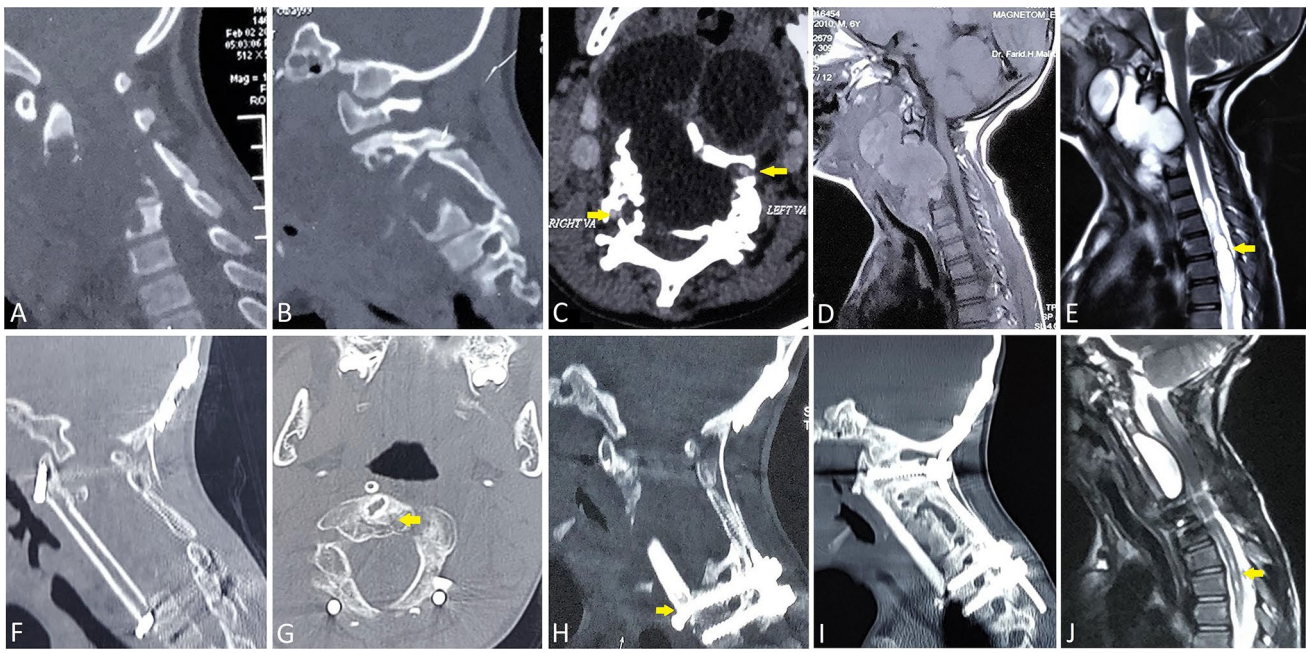

Fig. 1 Preoperative images (**a**–**e**): sagittal (**a**) and parasagittal (**b**) reconstruction of computed tomography (CT) scan show extensive osteolysis of the C2, fused C3–C5 and C6 vertebral bodies. **c** Axial CT section of subaxial spine (C3 level) shows vertebral body destruction; vertebral artery (VA) indicated with arrow. **d**, **e** On magnetic resonance imaging (MRI), the lesion is T1-isointense and T2-hyperintense and extends into retropharyngeal space. Syringomyelia is seen in the thoracic spine (arrow in **e**). **f**–**h** Postoperative CT at 1-year follow-up. Sagittal section (**f**) shows anterior column reconstruction using fibular strut graft with screws placed in the C1 anterior arch and C7; fusion is seen at graft–C1 interface (arrow in **g**) and at the caudal end (arrow in **h**). **i** Posterior occipito-cervical fusion was performed using occipital, C1 lateral mass, C7 and T1 pedicle screws). **j** Follow-up MRI shows resolution of syrinx (arrow)

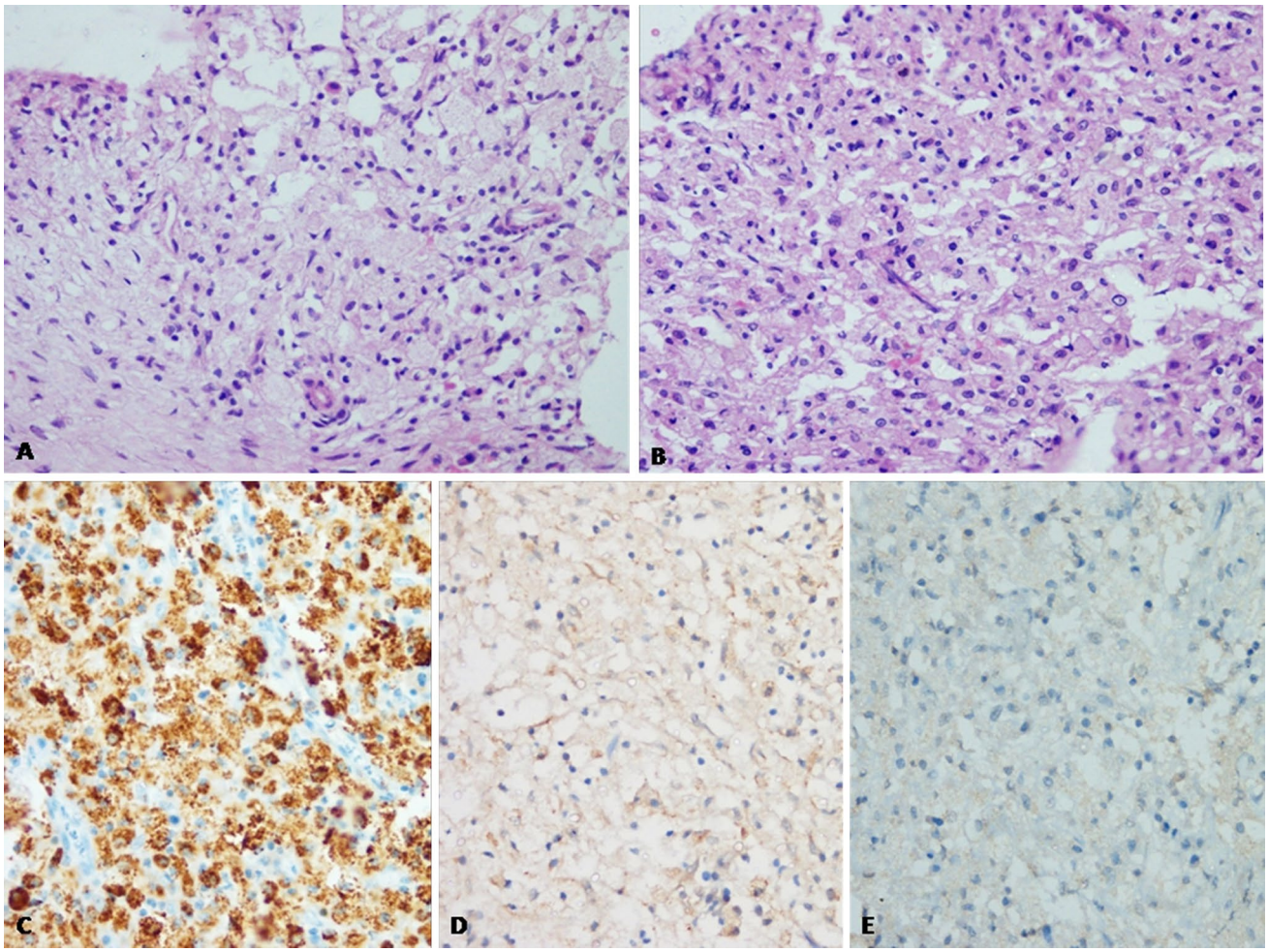

Fig. 2 **a** High magnification showing collection of foamy histiocytes and few lymphomononuclear cells. The infiltrate comprised chiefly of macrophages (H&E×400). **b–e** Foamy histiocytes and macrophages show strong and diffuse positivity with vimentin (**b**), CD68 (**c**) and are negative for S-100 (**d**) and CD1a (**e**) (**b–e**, immunoperoxidase ×400)

course [7]. The immunohistochemistry helps in this regard [1, 2].

JXG occurs primarily in the first two decades of life and most commonly (45–70%) within first 12 months of age. The most common presentation is a solitary dermal lesion (67%). Other less common presentations include solitary subcutaneous or deep soft tissue mass (16%), multiple cutaneous lesions (7%), solitary extracutaneous lesions (5%) and systemic disease (5%) [1, 2].

Solitary JXG involving the spine is unusual and can present in various forms such as an osteolytic lesion of the vertebral body, intradural or epidural mass, spinal nerve root lesion and rarely as an intramedullary tumor [3–6, 8]. Any part of the vertebral column can be involved. The MRI appearance varies from iso-hypointense in T1 and hypo-hyperintense in T2 sequence; the lesions usually show homogenous enhancement with contrast. Those lesions presenting as intradural extramedullary masses resemble meningioma on imaging, and the spinal nerve root involvement may mimic schwannoma [5]. In our patient, an extensive osteolytic lesion along with paraspinal and epidural spread made us think of possibilities such as spinal tuberculosis, aneurysmal bone cyst or osteolytic vertebral neoplasms.

The present case has certain unique characteristics. Firstly, the occurrence of solitary JXG in the region of craniovertebral junction (CVJ) is unusual. It was an extensive lesion involving almost the entire cervical spine (C2–C6) causing CVJ instability, and extending into the retropharyngeal space. Such long segmental disease involving multiple vertebral levels has not been reported. Secondly, there was a dilemma in the diagnosis because of its similarity to tuberculosis on imaging, intraoperative appearance and frozen sections. The presence of syringomyelia which was thought to be due to spinal arachnoiditis also supported a diagnosis of tuberculosis. Next interesting feature is the

association of the syrinx with an extradural lesion such as JXG. Though the reasons are unclear, it could be secondary to alteration of cerebrospinal fluid flow as in any other cervico-medullary lesion or due to an occult asymptomatic spinal arachnoiditis [9]. The syrinx resolution on follow-up MRI suggests that no additional procedures are necessary for the underlying syringomyelia in an extradural lesion.

Lastly, the extensive osteolysis caused by the tumor mandated a staged 360° fusion. In subaxial cervical spinal tumors with associated instability, the tumor excision is usually performed by an anterior approach, supplemented by mesh cage or strut graft. An additional posterior fusion may be required if the tumors extend to the lateral masses. In cases of solitary C2 body involvement, an anterior removal of the mass followed by an occipito-cervical fusion would suffice [10]. As the weight of the head is transmitted from the occipital condyle through C1–C2 lateral mass to the body of subaxial vertebrae, an additional anterior graft placement may not be necessary. However, in the event of associated destruction of C2 lateral mass or the body of subaxial spine, the major weight transmission would be borne by the posterior construct alone. This posterior construct is likely to fail over a period (cantilever effect), unless it is augmented by a load-sharing anterior strut (Fig. 3) [11].

Usually, an anterior extrapharyngeal approach is required to access the lesions that involve the C2 body along with multiple subaxial vertebrae. Reaching up to the clivus and obtaining its anchorage would be difficult through this approach and requires extensive dissection. In such cases, a strut graft can be placed between the intact C1 anterior arch and the uninvolved lower cervical spine, as described in the present report. We believe that ending the construct at C1 would provide an equally stable construct, thus obviating the need for an extensive dissection in a child. The autologous graft can be fashioned with ledges on the ends such that the body of the graft bears the weight, and also its cortical surfaces abut the anterior surface of C1 and the lower cervical spine (Fig. 3). A cortico-cancellous screw would firmly hold the graft until fusion occurs. Posteriorly, fusing the occipital squama to the C1 lateral mass and the subaxial spine is also necessary to prevent movement in between the occipito-atlantal joints that could dislodge the graft. Additionally, the C1 and the occiput act as a single unit, thereby sharing the weight transmission between the anterior strut graft and posterior construct.

A mesh cage or autologous graft is useful for an anterior column reconstruction. In our patient, a bone graft was preferred over a metallic cage as the latter could cause excessive sinking/subsidence in an immature bone; also a bone graft would allow some gain in the height of the vertebral column with time. Commonly, a rib graft is utilized in children for single- or two-level corpectomy [10]. This was not suitable in the present case due to a long-segment bony defect. Hence, we resorted to a fibular strut graft for the anterior column support. The upper screw was inserted through the fibular graft into the C1 anterior arch to stabilize the construct and prevent possible graft dislodgement. Such a technique can be a viable procedure to anchor the graft in cases with extensive C2 destruction. Although this has been occasionally described after C2 spondylectomy in adults, an insertion of tricortical screw anchored to the C1 arch in an immature spine has not been described to the best of our knowledge [12]. Furthermore, this technique is technically demanding in a pediatric spine due to a limited availability of C1 bone mass for screw purchase.

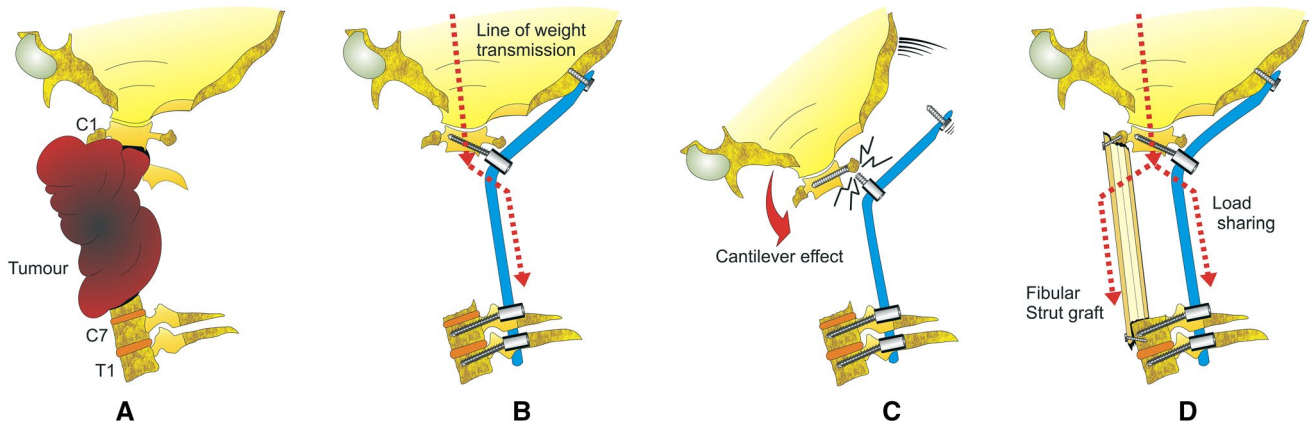

Fig. 3 Schematic diagram demonstrating the surgical technique and cantilever effect. **a** Osteolytic lesion extending from C2 to C6 vertebral bodies. **b** In the absence of an anterior graft, the entire weight of head is borne by the posterior occipito-cervical construct alone. The axis of weight transmission is indicated by dotted line. **c** Eventually, the posterior construct tends to fail due to cantilever mechanism (arrow). **d** The anterior strut graft allows load sharing between the anterior and posterior constructs and provides stable fixation. The fibular graft is fashioned with ledges on the ends such that the cortical surfaces abut the anterior surface of C1 and the lower cervical spine. The upper and lower screws are inserted through the graft into the anterior arch of C1 and the C7 vertebral body, respectively

Occasionally, cutaneous JXG has been reported in the context of neurofibromatosis type 1 [2]. The present case had an association with Klippel–Feil anomaly.

As far as the management of JXG is concerned, the majority of the classical skin lesions show spontaneous regression [2, 13]. In patients with systemic involvement, chemotherapy (steroids, vinblastine and methotrexate) is administered [1, 4, 13]. Owing to the rarity, no definitive treatment protocol exists for isolated spinal JXG. Total excision appears to be curative [3–6]. After partial excision, few have undergone adjuvant radiotherapy/chemotherapy [4, 14, 15]. Recently, neoadjuvant therapy using denosumab has been attempted in a pediatric patient [16]. A review of the reported cases with spinal involvement has shown an overall favorable clinical outcome with no recurrent disease [6]. Hence, we decided for an expectant treatment, considering the possible adverse effects of radiotherapy in young age.

Conclusion

Though uncommon, JXG should be thought of, in the differential diagnosis of solitary osteolytic lesions of vertebral bodies. They can simulate a tuberculous etiology on imaging and at surgery. At times, the lesions tend to be extensive and the management can be challenging. The described technique can be utilized for anterior column reconstruction in children with C2 body destruction.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Informed consent Informed consent was obtained from the patient.

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