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CASE REPORT

Primary Epidural Lumbar Ewing Sarcoma: Case Report and Review of the Literature

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Study Design. Case report.

Objective. We present a case of isolated primary epidural lumbar Ewing sarcoma and review the current literature on the standard management. We also propose laminoplasty as safe procedure in this patient population that can provide good stabilization in young people.

Summary of Background Data. Primary epidural Ewing's sarcoma is a very rare entity. The best generally accepted treatment option in sarcomas is to achieve a gross total resection with safe margins followed by local radiotherapy and chemotherapy. A total resection with safe margins is a great challenge in neurosurgical patients.

Methods. We present a previously healthy 17-year-old girl who complained of right sciatica with an epidural lumbar mass at L3-L4. She underwent complete resection of the tumor and a laminoplasty, which, in our experience, is a good way to preserve stability.

Results. At surgery, an isolated and noninvasive lesion was identified. Histopathological confirmation of Ewing sarcoma was obtained by immunohistochemical study and *EWSR1* gene rearrangement detection. Treatment with 6 months of chemotherapy resulted in no further identifiable lesions by PET and MRI imaging at 4 years postsurgery. The laminoplasty has remained stable.

Conclusion. Primary epidural Ewing sarcoma is extremely rare. The detection of the *EWSR1* gene rearrangement can help to diagnose these tumors. The decision on how to treat these patients is difficult and can hardly be based on data from the current literature because of the small number of patients. The laminoplasty procedure can be safely performed in the setting of sarcoma of the epidural space.

Key words: epidural, Ewing sarcoma, *EWSR1* gene rearrangement, laminoplasty.

Level of Evidence: 4

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Ewing sarcoma is a primitive neuroectodermal tumor originating in the medullar cavity of the diaphysis of long bones.[1] Initially, the histopathological diagnosis was based on the finding of blue small round cells on hematoxylin-eosin (HE) staining with no morphologic evidence of a cell of origin. Metastases usually affect lungs, long bones, and bone marrow. As a result, systemic chemotherapy has become an important component of treatment.[2]

CASE REPORT

A previous healthy 17-year-old girl consulted because of a 2 months' history of right sciatica. The neurological examination was normal.

Lumbosacral MRI showed an extradural $6'4 \times 1'8 \times 2$ cm mass at L3-L4 (Fig. 1). No other primary tumors or metastases were detected. After this finding the patient was proposed for surgery.

An L3-L4 laminoplasty was performed. At surgery we found an epidural isolated mass with no visible extension to adjacent bony structures. Complete resection was achieved.

Histopathological examination showed osseous, cartilaginous, and adipose tissue extensively infiltrated by a mitotically active small blue-round cell neoplasm suggestive of Ewing sarcoma (Fig. 2). Immunohistochemical profile is shown in Fig. 3. The Ki67 proliferative index was 40%, confirming the high mitotic rate. *EWSR1* gene rearrangement at the 22q12 locus was detected by fluorescence *in situ* hybridization (FISH) [LSI EWSR1 (22q12) Dual Color Break Apart (Vysis)], confirming the diagnosis.

After surgery, the patient was started on standard adjuvant combination chemotherapy. No radiotherapy was administered.

At 4 years of follow-up there have been no clinical or radiological evidences of local recurrence or metastases

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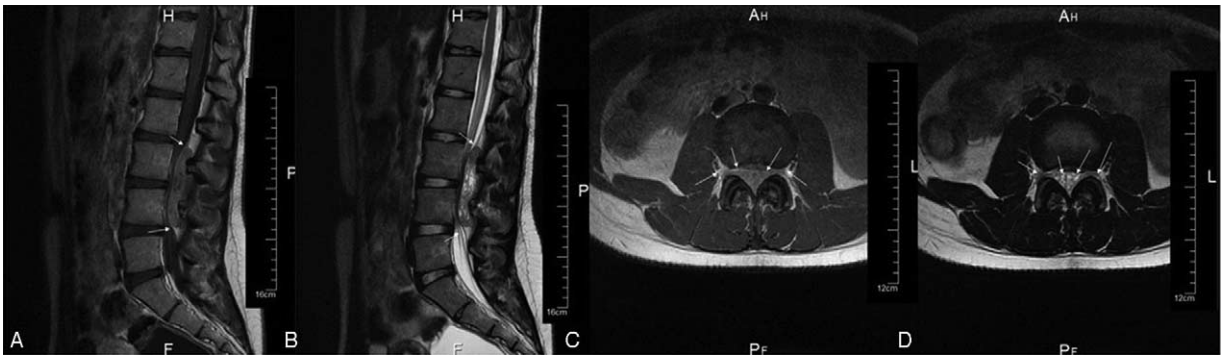

Figure 1. Sagittal and axial T1 with gadolinium (A and C) and T2 sequences (B and D) showing a dorsal epidural mass mainly on the right side at L3-L4.

(Fig. 4). The patient has also achieved a good stabilization of the spine.

DISCUSSION

Ewing sarcoma is the most frequent type of malignant bone tumor in children younger than the age of 10 years with the highest incidence in the second decade of life.[3]

Patients with Ewing sarcoma typically present with localized pain or swelling of a few weeks or months of duration. As Hsieh *et al*[4] published, around 70% of cases presented with neurological deficit, our patient did not. Machin *et al*[5] reported one case associated to 18q- syndrome, but it is the only case in the literature.

The best generally accepted treatment option in sarcomas is to achieve a gross total resection with safe margins, local radiotherapy, and chemotherapy. A total resection with safe margins is a major challenge in neurosurgical patients. Cotterill *et al*[6] and Rodríguez-Galindo *et al*[7] showed that patients with axial primary tumors had a worse outcome after treatment than patients with limb lesions. Kinsella *et al*[8] suggested that the key to management would be radiotherapy and chemotherapy and not surgery; nevertheless, nowadays a gross total en bloc resection has become essential for prognosis.

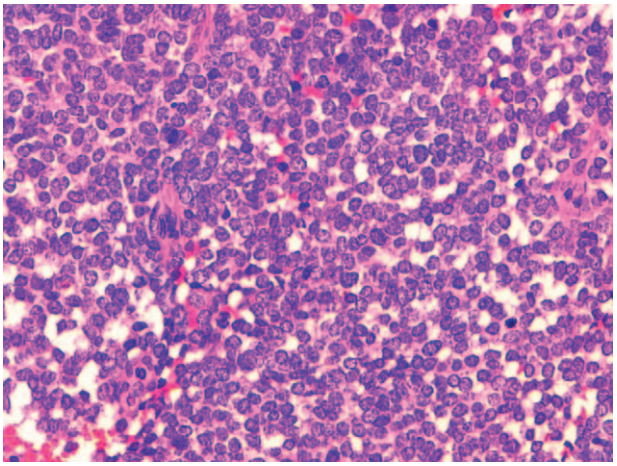

Figure 2. A highly cellular neoplasm composed of uniform round cells organized in solid/lobular pattern. Neoplastic cells showed round or ovoid nuclei with fine powdery chromatin and 1 or 2 small nucleoli and ill-defined, scant cytoplasm vacuolated in some (HE, 20×).

We consider that spinal radiotherapy should be reserved for residual lesions or tumors where surgery is not feasible. Kaspers *et al*[9] decided not to radiate their patient despite the incomplete resection basing their decision on the danger of neurologic sequelae resulting from damage to the spinal cord.

It is presumed that most patients will have subclinical metastatic disease at the time of diagnosis, even in the absence of overt metastases. Modern treatment plans all include chemotherapy.

Kaspers *et al*[9] performed a laminoplasty. This procedure can be questioned, but we have to bear in mind that this is an unusual tumor and the definitive diagnosis normally comes with the histopathological study after surgery. We performed a complete resection and a laminoplasty because, in our experience, this is a good way to preserve stability in young patients, there was a clear plane separating the tumor from the bone and the diagnosis of Ewing sarcoma was unexpected.

Hsieh *et al*[4] refers to prognosis according to place of origin, suggesting that the lumbosacral region might have a worse prognosis because of a later diagnosis. Our case is one of the few that, despite its lumbosacral origin, has had a good outcome. Kogawa *et al*[10] reported a cervical case with a good evolution after an early and aggressive management secondary to a quick onset of symptoms.

To our knowledge this is the only case in which *EWSR1* gene rearrangement has been used to establish the diagnosis. It is essential for rare tumors like this to have this type of ancillary techniques to confirm the diagnosis.

CONCLUSION

Children and young adults who present an epidural mass ought to be studied for a possible primary epidural Ewing sarcoma. The detection of *EWSR1* gene rearrangement can help to diagnose these tumors. The decision on how to treat these patients is difficult given the low number of cases.

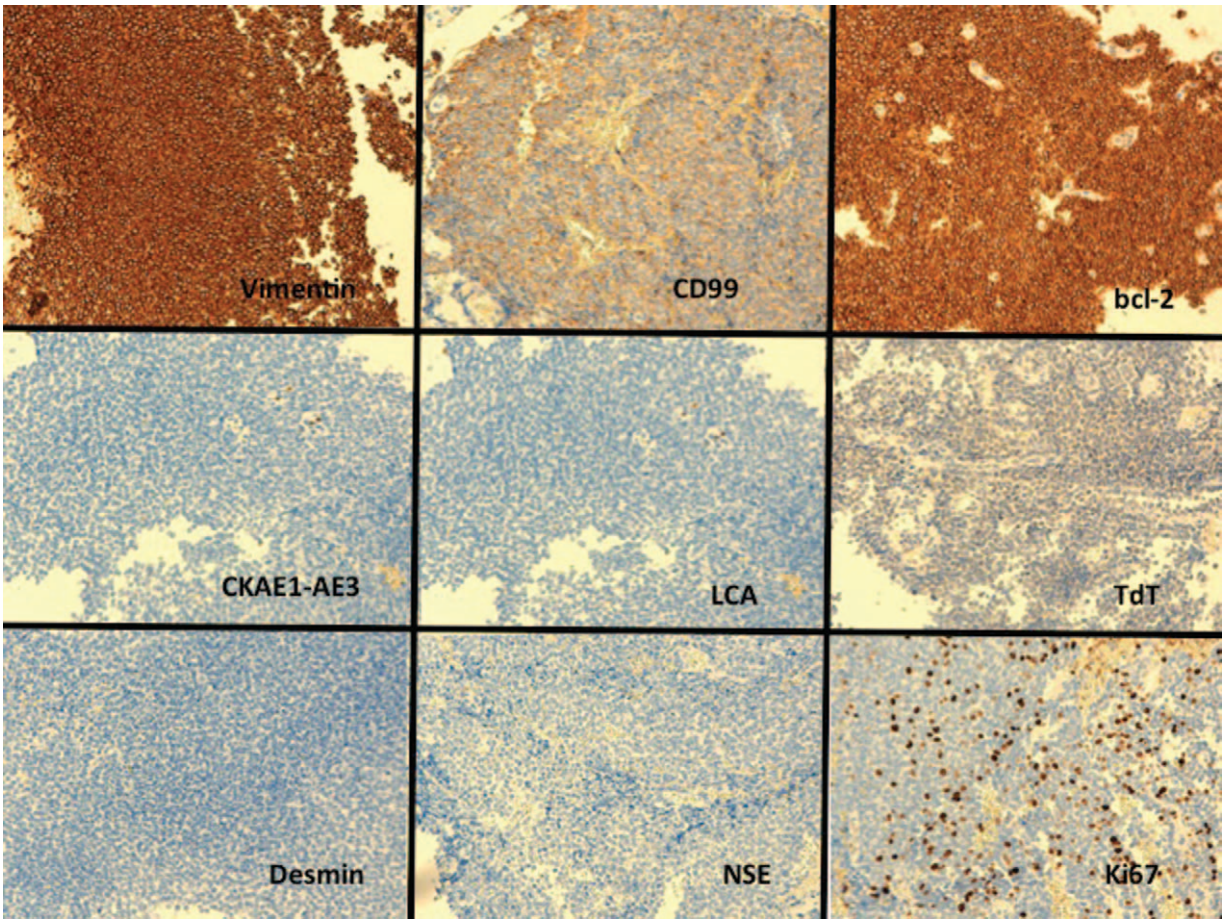

Figure 3. Immunohistochemically the neoplastic cells were positive with vimentin, CD99 and bcl-2 and negative with CKAE1-AE3, LCA (CD 45), TdT, Desmin, and NSE. The proliferative index (ki67) was 40% (IHQ, 20×). CK, cytokeratins; LCA, leukocyte common antigen; TdT, terminal deoxynucleotidyl transferase; NSE, neuron-specific enolase.

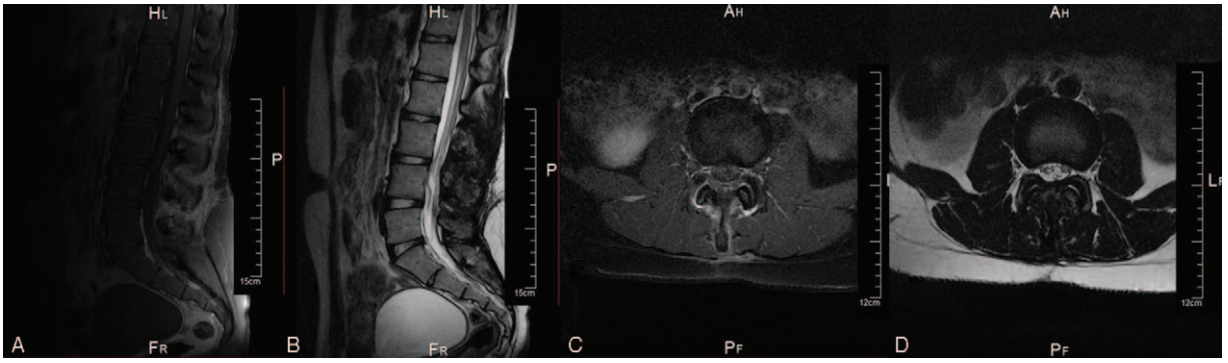

Figure 4. Sagittal and axial T1 with gadolinium (A and C) and T2 sequences (B and D) showing no residual tumor at 4 years follow-up.

➢ Key Points

- ❑ Primary epidural Ewing sarcoma with no bone affection is a very rare entity.
- ❑ Usual sarcoma management includes gross total resection with safe margins. This is a great challenge in neurological areas.
- ❑ Epidural Ewing sarcomas with a lumbosacral origin might have a worse prognosis because of a generally later diagnosis.
- ❑ The detection of the *EWSR1* gene rearrangement can help to confirm the diagnosis of these tumors.
- ❑ A laminoplasty procedure can be safely performed in the setting of sarcoma of the epidural space.

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