CASE REPORT

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Acute Paraplegia: A Rare Presentation of Askin Tumour

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Abstract

Background: Askin tumour is a rare, highly malignant chest wall neoplasm, most commonly observed in children and adolescents. It is a rapidly growing tumour with varied clinical manifestations and is associated with a poor prognosis. Given the non-specific clinical manifestations and aggressive nature of the disease, it is important to diagnose and treat early with a multi-disciplinary team involvement. In our case, a 14-year-old male presented with progressive left-sided chest pain and developed paraplegia due to compressive myelopathy, which is a rare presentation in this case. He was diagnosed with Askin tumour on histopathology and started on chemotherapy.

Key words: Askin tumour, paraplegia, compressive myelopathy.

Introduction

Askin tumour is a rare, aggressive malignant neoplasm of neuroectodermal origin, that arises from the soft tissues of the chest wall. It belongs to the Ewing sarcoma and primitive neuroectodermal tumour (PNET) family sharing histopathological characteristics of small round cells with variable degrees of neuroectodermal differentiation. The tumour occurs mainly in children and adolescents with a male preponderance (1.5:1). It is associated with a poor prognosis with a reported survival of 60% at 5 years[1,2,3].

The presentation can be variable, and symptoms range from being asymptomatic to prominent respiratory complaints such as chest pain, fever, cough, pleural effusions, and weight loss; making the diagnosis challenging[4]. Askin tumour causing acute paraplegia due to compressive myelopathy is extremely rare. Herein, we report a case of a 14-year-old male presenting with chest pain and acute paraparesis who was diagnosed with Askin tumour.

Case report

A 14-year-old male presented to our hospital emergency department with increasing left-sided chest pain for 20 days. He had no history of fever, cough, breathlessness. A chest X-ray showed left upper and mid-zone homogeneous rounded opacity (Fig. 1). A contrast computed tomography (CT) scan of the chest showed a large well-defined heterogeneously enhancing left apical mass lesion, eroding the posterior aspect of 2nd rib, measuring 8.6 x 7.2 x 9.3 cm (Fig. 2).

The patient was admitted to the ward for pain management and evaluation. His routine blood tests were within normal range apart from mild anaemia. A bronchoscopy and bronchoalveolar lavage (BAL) were performed from the left upper lobe. There was no endobronchial growth and BAL sample was negative for routine culture, acid-fast bacilli (AFB) smear, Gene-xpert and cytology. He developed sudden onset progressive bilateral lower limb weakness for 4 days and loss of sensation below the umbilicus and

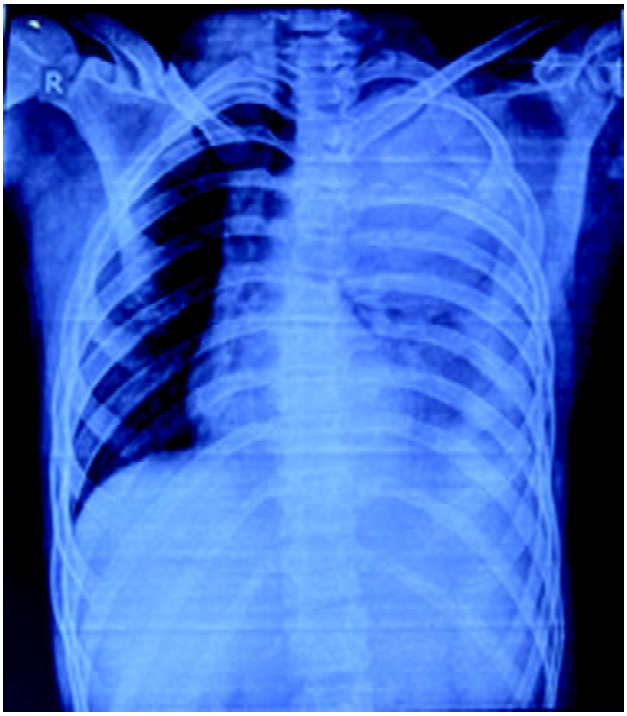

Fig. 1: *Chest X-ray showing round opacity in left upper and midzone.*

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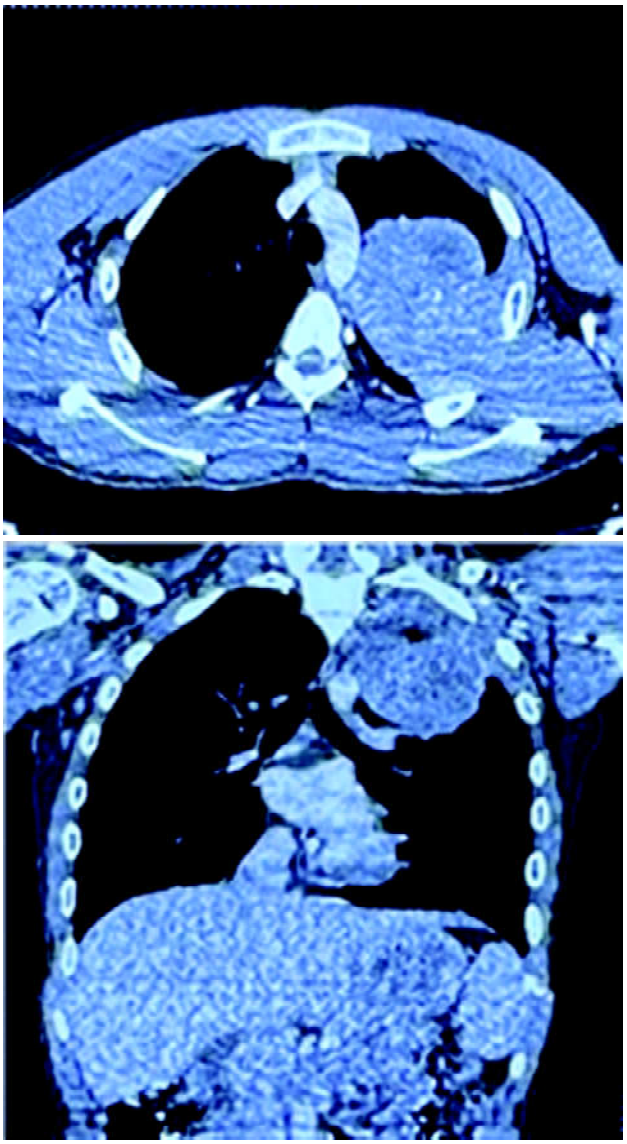

Fig. 2: *CT-chest demonstrating a heterogeneous left apical soft tissue mass eroding the posterior aspect of 2nd rib.*

urinary incontinence for 3 days. On neurological examination, his power was 5/5 in bilateral upper limbs and 0/5 in bilateral lower limbs along with brisk reflexes and loss of touch, vibration and joint position sense in bilateral lower limbs. Neurological consult was taken and the patient was started on intravenous methylprednisolone 500 mg, for suspected transverse myelitis. A magnetic resonance imaging (MRI) of the spine was performed that demonstrated a heterogeneous mass in the left upper thoracic cavity causing compressive myelopathy from D2 to D4, erosion of 2nd rib and infiltration into paraspinal muscles (Fig. 3).

Methylprednisolone was stopped and the patient was electively intubated for airway protection. Ultrasonography

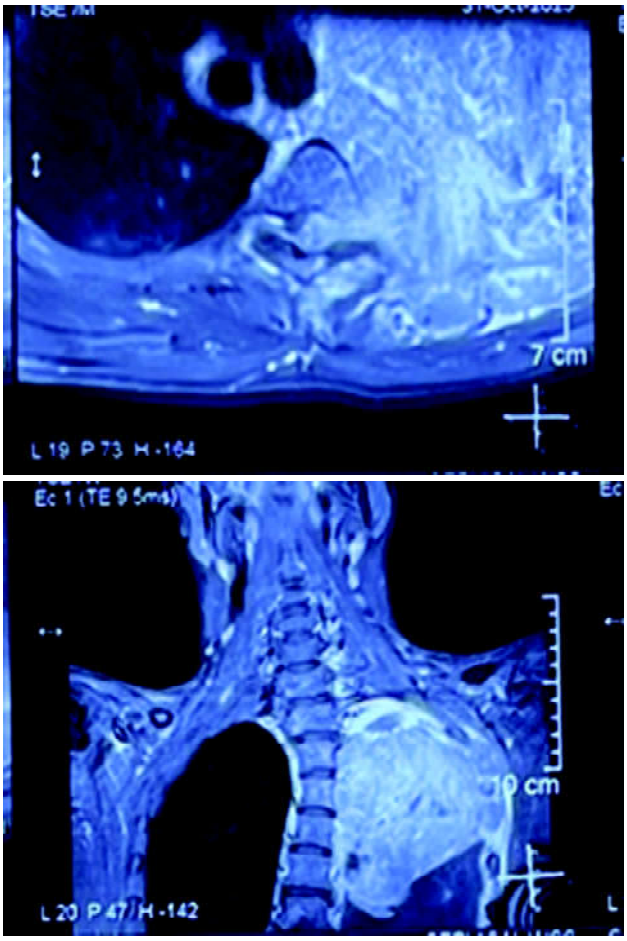

Fig. 3: *MRI showing heterogeneous hyperintense mass lesion in the left upper thoracic cavity causing compression from D2-D4.*

(USG)-guided tru-cut biopsy of the mass was performed via posterior approach. The histopathology reported the presence of a small round cell tumour (Fig. 4A). The immunohistochemistry was positive for CD99 suggestive of Askin tumour (Fig. 4B).

The patient was referred to oncology and initiated on chemotherapy with a Etoposide, Carboplatin and dexamethasone with a reassessment of tumour size after three cycles for radiotherapy and consideration for surgical resection.

Discussion

Askin tumours are small round blue cell tumours of soft tissues of chest wall that belong to the Ewing sarcoma family of tumours (ESFT). It is a highly malignant neoplasm with potential for loco-regional spread and recurrence, associated with poor prognosis. The clinical presentation can be non-specific including an incidentally detected mass on chest X-ray to a rapidly growing painful, palpable large mass[1,5]. However, a rapidly growing mass causing

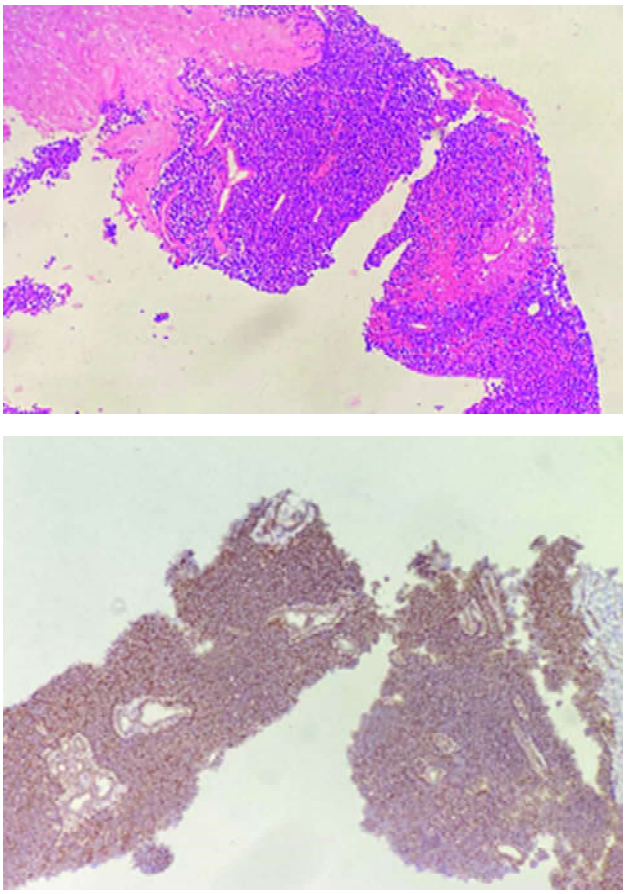

Fig. 4A: *Sheets of round to oval cells with* ***(B):*** *CD99-strong membranous positivity scant cytoplasm.*

compressive myelopathy and acute paraparesis are rare presentations, and to our knowledge, only two cases in the literature have been reported so far. The differential diagnosis that was considered in this case were acute transverse myelitis, and Gullian Barre syndrome (GBS).

These tumours appear as heterogeneous masses on MRI and CT scan with areas of cystic degeneration, necrosis, haemorrhage and pleural effusions. Rib destruction is observed in 25 - 63% of cases[6].

Primary chest wall tumours constitute less than five per cent of thoracic neoplasms, with a malignancy rate of fifty per cent. Fifty-five per cent arise from bone or cartilage and forty-five per cent arise from soft tissues. About twenty per cent are discovered incidentally. The most common primary chest wall malignant tumour is chondrosarcoma; other differential causes include rhabdomyosarcoma, plasmacytoma, neuroblastoma, lymphoma, neurofibroma and metastatic disease[4,7]. Our case was diagnosed based on the aggressive nature of the disease, biopsy findings of small round blue cells and positive CD 99 stain on IHC.

There are limited clinical trials to provide a definite consensus on management. A multimodality treatment approach of chemotherapy followed by *en bloc* resection and adjuvant chemoradiation is associated with a 5-year overall survival rate of 60.7%. Poor prognostic factors include age >18 years, poor response to induction chemotherapy, and presence of pleural effusion[8].

Conclusion

Askin tumour is a rare cause of a chest wall mass with variable and non-specific clinico-radiological presentation. It is an aggressive tumour that can cause acute paraplegia due to compressive myelopathy and should be kept as a differential in a young patient with a chest wall mass presenting with paraparesis. Hence, timely diagnosis and prompt multidisciplinary team involvement can prolong the overall survival significantly.

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