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Case Report

A case of Askin's tumor presenting as acute paraplegia

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ABSTRACT

Askin tumor is an uncommon malignant neoplasm of a neuroectodermic origin that arises from the soft tissues of the thoracopulmonary wall. Defined histologically by malignant small round cell tumor and immunohistochemistry positive for CD99. The lack of clinical guides that establish a standardized management contributes to its poor prognosis and short overall survival.

1. Introduction

Extra Osseous Ewing Sarcoma is a rare aggressive malignant tumor mainly affecting young people of age group between 5 to 35 years of age. Askin tumor is rare type Ewings sarcoma at thoraco-pulmonary region.[1] It has got high mortality rate. Diagnosis is based on imaging modalities followed by histopathological examination. Early diagnosis is essential for better outcome. We report a case of 4 yrs old boy with extra skeletal Ewing sarcoma of thoracic spinal cord. The patient underwent hemilaminectomy followed by chemotherapy and radiotherapy.

2. Case Report

A four years old boy, developmentally normal, immunized for age presented with, cough for 1 week, weakness of both lower limb for 4 days and breathlessness for 1 day. Child had nonproductive cough for the past 1 week. Weakness was initially noted in his right lower limb and after one day involved both lower limbs. It was progressed with in 4 days making him unable to walk. On fourth day of the weakness child developed breathlessness which was not relieving with medications. Child also lost bladder sensation within 1 week of hospital admission.

Child is having loss of weight and appetite for the past 2 weeks. No history of fever, altered sensorium, seizures or trauma. No history of any sensory symptoms. No history suggestive of weakness of upper limbs or cranial nerve involvement. No history of recent vaccination. No family history of tuberculosis or any other significant history in the past.

On examination child was irritable. Lying on bed with both lower limbs extended and externally rotated. He was tachypneic but afebrile. His anthropometric measurement showed grade 2 PEM.

On nervous system examination, child was conscious and oriented. Cranial nerve examination was normal. Noted to have hypotonia of both lower limbs with a power of grade 2 on both lower limbs. Abdominal reflex and cremasteric reflex were absent with bilateral extensor plantar response. Deep tendon jerks were absent. Sensory function decreased below the level of T6 vertebrae. Cerebellar signs were negative. No signs of meningeal irritation.

On respiratory system examination, noted to have decreased chest movements over right side. Fullness of chest

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with bulging intercostal spaces. Trachea shifted to left side. Stony dullness noted over right side on percussion. Air entry was decreased over right side.

On investigation his Hb was 10.9gm% and TC 11,700cells/mm3. Chest Xray showed massive pleural effusion on right side with tracheal shift (Figures 1 and 2). Intercostal chest drain was inserted. Pleural fluid study showed 20 cells with 3g protein and normal sugar. It was transudative effusion. No atypical cells seen. No organism seen in gram stain. CBNAAT negative and culture was sterile.

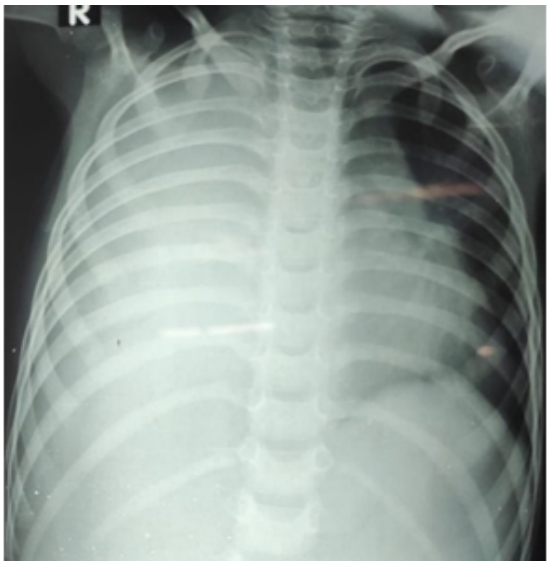

Figure 1: Chest Xray showing right sided pleural effusion

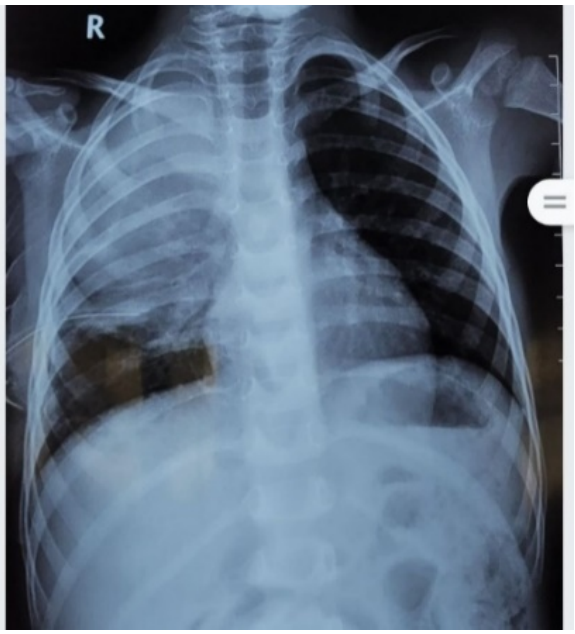

Figure 2: Chest Xray after intercostals tube insertion and drainage.

CSF study showed showed high protein (albumin cytological dissociation). Nerve conduction study was normal. IVIG (2g/kg) was given suspecting GBS. Anti-tuberculosis treatment was started suspecting pulmonary tuberculosis. In view of his persistent weakness and bladder involvement MRI brain and spine was done. MRI showed extradural extramedullary fairy well defined lesion involving dorsal spine extending from D1 to D6 vertebral bodies with extraspinal extension into the right paraspinal extra-pleural space through right D3-D4 and D4-D5 neural foramens (Figures 3 and 4).

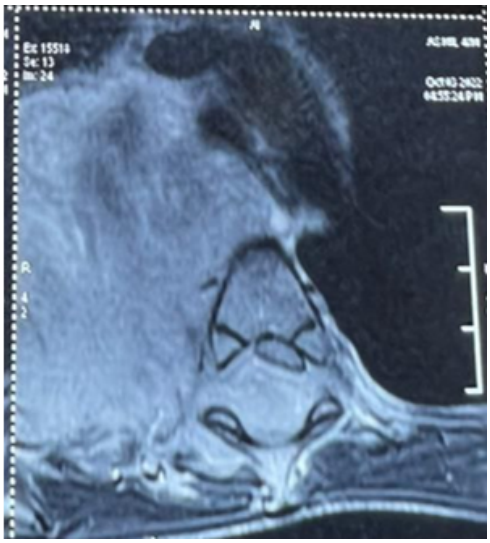

Figure 3: MRI showing extension of mass to posterior and superior mediastinal space.

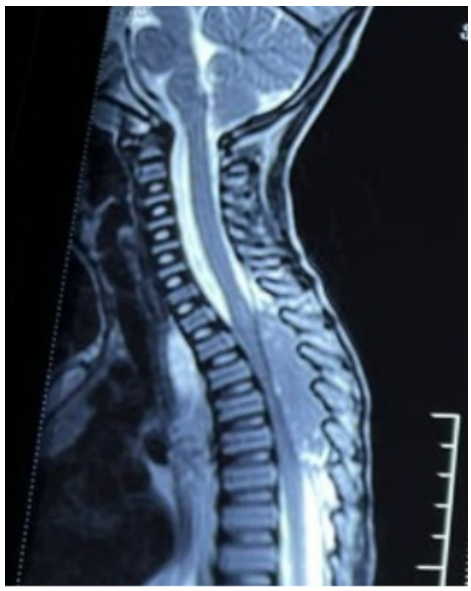

Figure 4: MRI showed extradura extra medullary lesion extending from D1 to D6 spinal cord segments.

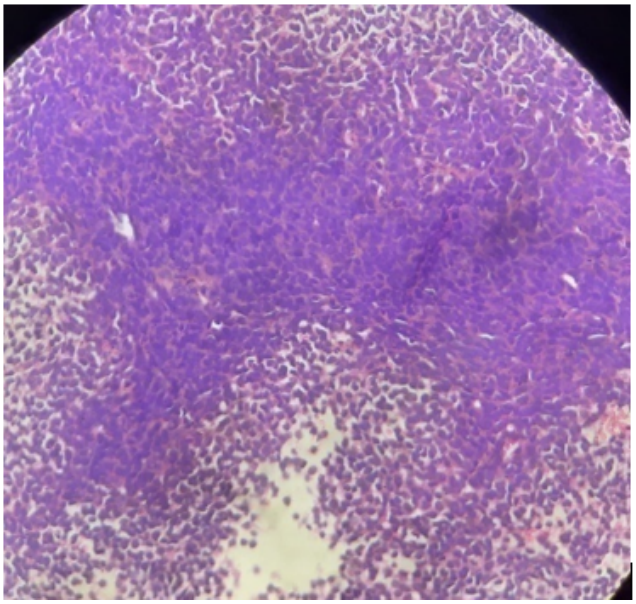

Figure 5: Histopathology showing uniform small round cells, finely stippled chromatin, inconspicuous nucleoli, eosinophilic cytoplasm, sheet-like growth pattern suggestive of Ewings sarcoma

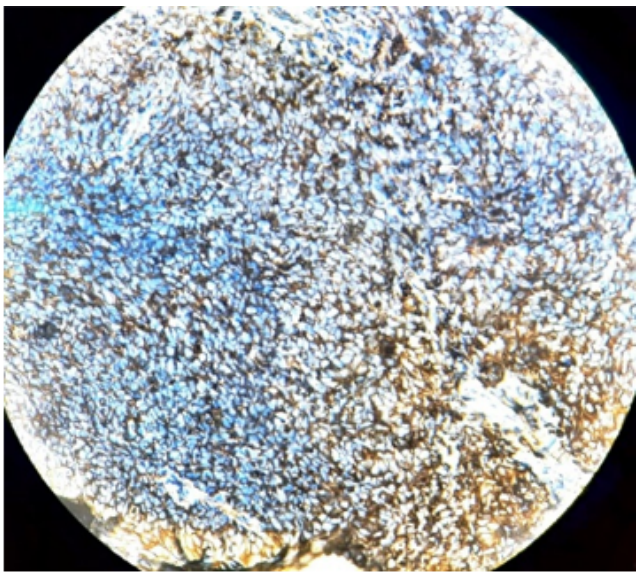

Figure 6: Immunohistochemistry showing CD99 positive

Child underwent D1-D4 right hemilaminectomy and tumor excision from thecal sac was done. Histopathological study showed uniform small round cells 1 - 2x size of lymphocytes with round nuclei, finely stippled chromatin, inconspicuous nucleoli, scant clear to eosinophilic cytoplasm, indistinct cytoplasmic membranes, sheet-like growth pattern, islands separated by dense fibrous tissue. Immunohistochemistry was strongly positive for CD99 which was suggestive of extraosseous Ewing sarcoma (Figures 5 and 6).

After surgery child's symptoms were persisting. Antituberculous treatment stopped. He was referred to higher center for chemotherapy and further management. Bone scan was normal.FISH probe assay done which showed chromosomal rearrangement involving EWSR1 gene on chromosome 22 q 12. He was started on chemotherapy followed by radiotherapy. Now child is improving and able to stand without support.

3. Discussion

Askin tumors(AT) belong to Ewing family of tumors with a variable degree of neuroectoderm differentiation.[1] Other thrtee subsets of tumors in Ewing sarcoma family of tumors (ESFT) include Ewing sarcoma of the bone, peripheral primitive neuroectodermal tumor (pPNET), and the extraosseous or extraskeletal Ewing sarcoma (EES).

Histologically, AT show small round blue cells. They originate from thoracopulmonary region. First described for the first time in 1979.[1] Askin tumor presents with respiratory problems such as chest pain, dyspnea and weight loss. It is highly malignant with poor prognosis and short survival. The reported overall survival is 60% at 5 years.[2,3]

The present case presented with weakness of both lowerlimb, a rare type of presentation in a four-year-old child. Later on investigation found to have right sided thoracic mass with right sided pleural effusion. Diagnosis of AT was established based on the features like- aggressive nature of presentation and biopsy findings (uniform small round cells, finely stippled chromatin, inconspicuous nucleoli, eosinophilic cytoplasm, sheet-like growth pattern with CD99 positivity).

Poor prognostic factors of AT include: age >18 years, poor response to induction chemotherapy, and presence of pleural effusion.[4]

Because of rarity of condition, there is no defined treatment guideline for this condition. Most centers follow multimodality treatment of chemotherapy, surgery, and radiotherapy.

There are only few case reports of AT from India The case is presented because of its rarity, atypical clinical presentation and imaging findings[5–7] Askin tumor presenting as compressive myelopathy is extremely rare. The common differential diagnosis acute flaccid paralysis in children include GBS, transverse myelitis and traumatic neuritis. Very rarely we have to keep the possibilities like Askins tumor in a case of paraplegia.

4. Source of Funding

None.

5. Conflict of Interest

None.

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