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INTERESTING MEDICAL IMAGE

Askin's Tumour

Massive tumour with minimal symptoms

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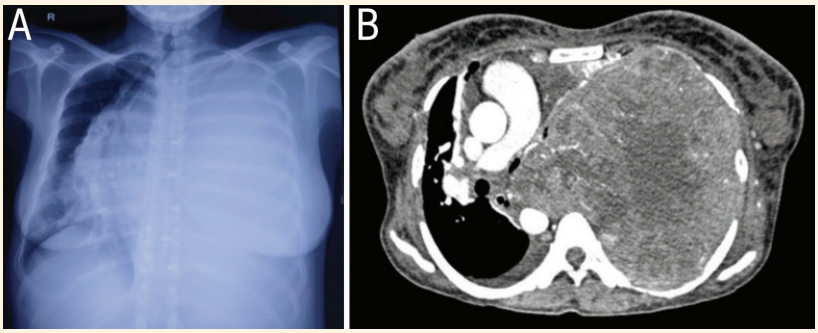

Figure 1: A: Chest radiography showing the left hemithorax (whiteout) and gross contralateral shifting of mediastinal structures and **B:** Computerised tomography of the thorax (mediastinal window) at level T-5 showing a huge mass lesion virtually replacing the left lung and gross contralateral shifting of mediastinum in a 33-years-old female patient with Askin's tumour. The mass lesion also shows various necrotic areas interspersed in between.

A 33-year-old female patient presented to a tertiary care hospital Bhopal, India, in 2019 with complaints of left-sided chest discomfort for four months and progressively increasing shortness of breath for one month. The patient developed hoarseness of voice, chest pain and dysphagia one week before presenting to this hospital. The chest X-ray revealed a large homogenous opacity in the left hemithorax with tracheal and cardiac deviation to the right side [Figure 1A]. A contrast enhanced computed tomography of the thorax revealed a single large heterogeneously enhancing soft tissue mass lesion occupying the entire left hemithorax, measuring 27 × 14.6 × 15.5 cm [Figure 1B]. The lesion had caused a significant mass effect in the form of a gross contralateral mediastinal shift, inferior displacement of the diaphragm, anterior displacement of the spleen, encasement and obliteration of the left pulmonary artery and abutment and displacement of the left subclavian artery, arch of aorta and descending aorta without any luminal narrowing. No obvious bony lytic or sclerotic lesion was noted.

An ultrasonography-guided fine needle aspiration cytology of the lesion revealed a malignant round cell tumour, potentially qualifying as a primitive neuroectodermal tumour (PNET). A biopsy of the lesion revealed malignant round cell tumour [Figure 2A]. Immunohistochemistry of the lesion was positive for cluster of differentiation (CD)99 [Figure 2B]. The lymphoid markers—*Wilms' tumour 1* gene and epithelial membrane antigen—were negative. Apart from CD99 positivity, the characteristic site and morphology of the tumour supported the diagnosis of PNET. No blasts were observed on peripheral blood and bone marrow examination, excluding the possibility of lymphoblastic malignancy. There was also no sign of desmoplasia in the tumour stroma thereby excluding the diagnosis of a desmoplastic small round cell tumour. The patient was started on chemotherapy with a VAC-IE (vincristine, adriamycin, cyclophosphamide followed by ifosfamide and etoposide) regimen on the advice of a multi-disciplinary team. Serum lactate dehydrogenase (LDH) level was at 904 U/L. A bone marrow aspiration and biopsy done for staging purposes revealed hyper cellular marrow with erythroid hyperplasia and mild megalobastosis with no evidence of marrow infiltration by tumour cells. The patient's performance status as per the Eastern Cooperative Oncology Group scale was 3/5. She received her first cycle of chemotherapy as per protocol and supportive treatment in the form of feeding through a nasogastric tube, analgesics and hydration. On the fourth day post-chemotherapy, the patient developed hypoxemic respiratory failure which was managed with supplemental oxygen and antibiotics. However, her condition worsened over the next few days with the patient succumbing to respiratory failure.

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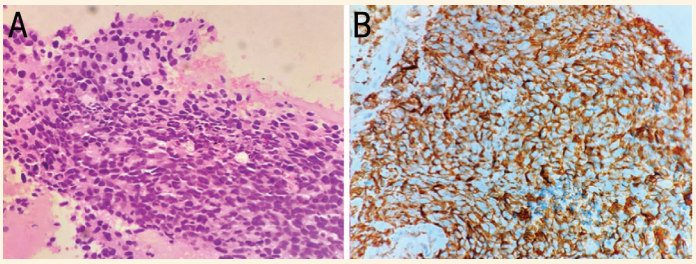

Figure 2: Ultrasonography-guided biopsy showing a hypercellular tumour lying in sheets composed of small, round, hyperchromatic tumour cells with scant cytoplasm in a 33-year-old female patient with Askin's tumour. **A:** Haematoxylin and eosin stain at ×200 magnification showing tumour cells that are seen encircling lumen forming rosettes at certain places. **B:** Immunostaining with cluster of differentiation 99 at ×200 magnification showing tumour cells displaying strong membranous positivity.

Consent was obtained from the patient's husband to utilise her case record and images for publication purposes.

Comment

PNET of the thoracopulmonary region, also referred to as Askin's tumour, was first described in 1979 by Askin *et al*. and is a very rare entity.[1] Developing from the soft tissues of the chest wall, particularly in the paravertebral region, the clinical symptoms include painful chest wall mass and other associated symptoms such as dyspnoea, cough, weight loss and regional lymphadenopathy. A chest wall soft-tissue density mass, sometimes associated with rib erosion and/or pleural effusion, is the most commonly found radiographic manifestation. It may only produce vague discomfort occasionally, thereby delaying the diagnosis to a very advanced stage.[2] The differential diagnosis of such a large unilateral soft tissue density includes Ewing's sarcoma, rhabdomyosarcoma, neuroblastoma and ganglioneuroblastoma.[2,3]

To the best of the authors' knowledge, the current case of Askin's tumour is the largest tumour reported in existing literature (27 × 14.6 × 15.5cm) compared to the previously reported maximum of 15 × 22.5 cm.[4] The largest tumour in a case series of 20 patients reported by Askin *et al*. had a maximum diameter of 14 cm.[1] The maximum size of Askin's tumour in a series of 11 patients reported by Zhang *et al*. was 12.5 cm and was 13.7 cm in another series reported by Xia *et al*.[3,5] The relatively stable vitals at admission and lack of rib erosion and pleural effusion, for a mass of this size, were quite unique to this case.

The histopathology of Askin's tumour typically reveals undifferentiated sarcomatous tissue of small round cells with scant eosinophilic cytoplasm, high nuclear-cytoplasmic ratio, small single nucleoli and a high mitotic rate.[6] Immunohistochemistry is generally positive for CD99 and neuron specific enolase. Presently, the diagnosis of Askin's tumour is based on oncogenetic practices that look for chromosomal translocations (t [11;22][q24; q12]) and/or their fusion transcripts. This mutation is also seen in Ewing's sarcoma.[6] The prognosis of this tumour remains poor due to local recurrence and distant metastasis, usually to the lungs and skeletal system.[3] The factors determining prognosis include size of the primary tumour, LDH levels, distant metastasis and performance score of the patient.[4] These factors predicted a poor prognosis in our patient who died shortly after starting chemotherapy.

AUTHORS' CONTRIBUTION

AH, AKK and AG were involved in patient management. AH and AKK performed the literature review. AH drafted the manuscript, while AKK, AG and DJ edited the manuscript. DJ performed the laboratory diagnosis. All authors approved the final version of the manuscript.

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