Alveolar soft tissue sarcoma: rare presentation of a rarer malignancy

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A 60-year-old postmenopausal female presented with complaints of a rapidly growing ulcerated proliferative growth in the left supraclavicular fossa since last 4 months. The mass lesion started in the supraclavicular fossa left side and rapidly extended downwards infiltrating over the anterior chest wall (Fig. 1). Patient gives no history of pain over the mass, cough or chest pain, restriction of movement of neck and shoulder, parasthesia in the left arm and forearm, upper limb claudication or edema.

On examination of the mass, it was found to be 15 x 10 cm, globular, extending over the left supraclavicular fossa upto the shoulder joint laterally and inferiorly over the anterior chest wall and medially till the root of the neck. It was hard in consistency, non tender, bled on touch, irregular margins, fixed to the clavicle and chest wall, with no induration over the surrounding skin. Ranges of movements of the left shoulder joint were not restricted. No edema, wasting or signs of vascular insufficiency in the left upper limb with no neurological deficits.

Other systemic examinations were within normal limits.

Provisional differential diagnosis of soft tissue sarcoma and carcinoma of accessory breast was made and patient was

subjected to routine blood investigations and an incisional biopsy, which reported it to be alveolar soft part sarcoma (Fig. 2). Immunohistochemistry stains revealed positivity to Desmin and PAS stains. Radiologically soft tissue shadow was seen with no bony abnormality and clear lung fields.

Alveolar soft part sarcoma is classified amongst the malignant and highly aggressive soft tissue sarcomas [1] carrying a poor prognosis. The usual age of presentation is 15 to 35 years with female preponderance under the age of 20 years [2]. It is a rare tumor with a predilection for the lower extremities in the adults and the head and neck in children. It is composed of large cells with granular eosinophilic cytoplasm. These cells are arranged in nodules, which are separated by each other by thin walled vascular channels.

Fig. 1 Showing mass lesion

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Fig. 2 Showing Alveolar soft part sarcoma, found by routine blood investigations and an incisional biopsy.

There is no clear scientific evidence as to the histogenesis of this tumor with some data supporting neural derivation, while others suggest myogenic origin [3].

It is a slow growing tumor with a high propensity for vascular spread and early distant metastasis. It has a predilection to metastasize to lung, brain and bone. Patients with metastatic disease may remain asymptomatic for years with the slow growing primary, the ultimate prognosis is poor.

References

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