Case Report

Metatastatic Alveolar Soft Part Sarcoma---A Case Report

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Introduction

Alveolar soft part sarcoma is a rare neoplasm, usually seen in young patients of < 30 years of age. There is an age range of 0-74 years. Most of the patients are young females. It usually occurs in extremities specially thigh, buttock, oral cavity, pharynx including tongue, mediastinum, sometimes from pulmonary vein, stomach, retroperitonium, uterus, vagina, and orbit. It is a very vascular tumour and bruit is usually present. Local recurrence rate is 20-33%. This neoplasm is known to have late recurrences. Incidence of metastasis is 66%.

Here we report a case of alveolar soft part sarcoma metastasizing to the brain where the patient presented as a space occupying lesion (SOL). After a histopathology diagnosis of metastatic tumour, a search was made for primary lesion which culminated in a biopsy of thigh swelling revealing alveolar soft part sarcoma.

Case Report

A 33 year old female resident of Nowshera, presented in Neurosurgery Department, Pakistan Institute of Medical Sciences on 25-4-2005 with complaints of left sided hemiplegia, headache off and on, vomiting, loss of vision in left eye, reduced vision in the right eye and history of weight loss for the past three months. There was no history of fits, fever or any other systemic manifestations. A swelling on right upper leg, inner aspect was operated some ten years back. There was no other significant, family or drug history.

On examination she was found to have stable vitals. Her central nervous system examination revealed intact higher mental functions, with a GCS of 15/15. On eye examination the right eye was deviated to the left and pupils of both the eyes were fixed and non reactive to light. There was 6th nerve palsy. On examination of the motor system the positive finding was that, the power in the left upper limb was reduced to 2/5. Tone and reflexes were reduced in the same limb. Plantars were bilaterally down going. Rest of the systemic examination was unremarkable. There was a swelling on the inner aspect of right upper leg measuring 7x 4 cm. The temperature of the swelling appeared to be hot.

On investigations her labs were unremarkable. CT scan of the brain with and without contrast revealed a mixed density SOL at the parietal region.

On the basis of history, examination and investigations a differential diagnosis of high grade glioma, tuberculoma & metastatic lesion were considered.

Craniotomy was done and peroperatively the tumour was found to be vascular, soft to firm, well circumscribed, and suckable. It was removed piecemeal and sent for histopathological examination. The patient was stable after the operation but there was no improvement in the neurological deficits. The biopsy of the swelling on her leg was performed on 13.4.05 and then she was discharged three days later after giving her oral medications including, tab Vixime, tab Epival, tab Hyocid, tab Decadron and tablet Dijex.

Material and Methods

The specimen was fixed in 10% buffered formalin. The tissue was routinely processed and stained with hematoxylin and eosin. PAS stain was performed on additional sections.
Pathological Findings

Grossly the tissue consisted of multiple soft tissue fragments grayish brown in colour weighing 13 Gms. Representative sections were submitted

Then we received a biopsy from the swelling in the leg of the patient, which was grossly a soft tissue piece 0.8 x 0.5 cm in size. Microscopic examination revealed the same malignant neoplasm with a few solid nests at the periphery of the section. The vessels in the septae were infiltrated by the tumour cells

The PAS stained section revealed PAS positive diastase resistant material in the cytoplasm of the neoplastic cells. On the basis of these findings a diagnosis of alveolar soft part sarcoma metastasizing to the brain was made

Comments and Discussion

Alveolar soft part sarcoma is a malignant neoplasm designated in the past as organoid granular cell myoblastoma and malignant non chromaffin paraganglioma. It is seen in the younger age of < 30 years, with an age range of 0-74 years. Most of the patients are young females. It usually occurs in extremities specially thigh, buttock, oral cavity, pharynx, including tongue, mediastinum, sometimes from pulmonary vein, stomach, retroperitonium, uterus, vagina, and orbit.¹, ² This tumour is very vascular and bruit is usually present. Local recurrence rate is 20-33 %. It is known to have late recurrences.³, ⁴ Incidence of metastasis is 66 %. The five years survival is quoted to be 59-67 % and 10 years survival is 47%. Most of the patients eventually die of the disease.¹, ²

Grossly the tumours are well circumscribed, usually large, moderately firm and greyish or yellowish. Areas of hemorrhage or necrosis are common in large neoplasms. No other sarcoma has quite the same appearance as alveolar soft part sarcoma. The majority of lesions contain large areas with an alveolar pattern, which is well illustrated by a reticulin stain, surrounding nests rather than individual cells. The cells themselves are uniform and have large round nuclei with a single prominent nucleus, together with granular eosinophilic or clear cytoplasm PAS-positive, diastase-resistant crystalline material (apparently actin) can be found in most cases, but it is quite variable in quantity. A common feature is vascular invasion. As a rule, mitoses are scarce. Unusual variants without much alveolar formation and scattered pleomorphic cells have been reported Alveolar soft part sarcomas occurring in children are often associated with a more solid pattern of growth and tend to be misdiagnosed.¹

The differential diagnosis includes paraganglioma, rhabdomyosarcoma, renal cell carcinoma, metastatic adrenal carcinoma, clear cell carcinoma and melanoma. Unlike paragangliomas, the cells are frequently discohesive and lack nuclear pleomorphism. Unlike alveolar RMS, the alveoli are not elongated and are not separated by fibrous tissue, and the cells are much larger than primitive myocytes. The tumor may closely resemble areas in renal cell carcinoma, melanoma, metastatic adrenal carcinoma (often
pleomorphic), and clear-cell sarcoma. The natural history can be quite prolonged.\(^1\)

Ultra structurally the cytoplasmic crystals are membrane bound crystals with a periodicity of 58 to 100 nm, sometimes arranged in a cross grid pattern. This feature is of great diagnostic value in lesions of controversial nature. Other ultra structural features include vesicles with an electron dense content in the golgi region, which possibly represent the precursors of the crystals, and smooth tubular aggregates associated with plasmalem invaginations.\(^5\)

The neoplasm has been known to be associated with chromosomal unbalanced translocations der (17) t (x; 17) (p11.2; q25), which results in the fusion of the TFE 3 transcription factor gene to the ASPL gene. The presence of aberrant nuclear expression of TFE 3 can be demonstrated in these tumours by immunohistochemistry.\(^1\)

The histogenesis of this tumor has been argued ever since its description in 1952. Recently, interest has centered upon a myogenic origin as a result of the finding of desmin, actin, and other muscle markers. Several examples with both desmin and MSA in contrast to previous results are seen. Staining is never extensive. A skeletal muscle origin seemed further supported by the detection of cytoplasmic MyoD1 protein, but more specific MyoD1 nuclear staining is absent. Whether this unusual lesion derives from a specialized muscle cell or other cell type still remains an open question. No one has yet reported the presence of myoglobin.\(^1\)

The Japanese musculoskeletal oncology group studied 57 patients of alveolar soft part sarcoma for 20 years in subjects with age range of 7-75 years. Of these there were 37 females and 20 males. Out of these 13 showed bone involvement at primary site and 6 of them were diagnosed as bone tumours at presentation. Tumour size, bone involvement at the primary site and presence of metastases at presentation were prognostic indicators (P< .05).\(^6\)

Another study which reviewed CNS involvement in children affected by sarcomas in Israel revealed 19 cases of CNS metastases of which one was that of alveolar soft part sarcoma.\(^7\)

References