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METASTATIC ALVEOLAR SOFT PART SARCOMA IN BRAIN, AN INCIDENTAL DIAGNOSIS IN AUTOPSY DEPARTMENT OF PATHOLOGY: A CASE REPORT

Pathology

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ABSTRACT

Alveolar soft part sarcoma (ASPS) is a rare malignant soft tissue tumor accounting for approximately 0.4–1% of all soft tissue sarcomas. It predominantly affects young adults, with a female preponderance, and commonly arises in the deep soft tissues of the thigh or buttock. The tumor is characteristically slow growing and painless, often leading to delayed diagnosis, with metastasis as the initial presentation. Among sarcomas, ASPS has a notably high predilection for brain metastasis. We report an autopsy case of intracranial metastasis from ASPS in a young adult, highlighting its clinicopathological features and diagnostic challenges.

KEYWORDS

Alveolar Soft Part Sarcoma, Metastasis, Autopsy, Soft Tissue

INTRODUCTION

Alveolar soft part sarcoma is an uncommon malignant mesenchymal tumor characterized by a distinct alveolar growth pattern and unique molecular alterations. It primarily affects adolescents and young adults between 15 and 35 years of age, with a female predominance. The most frequent primary site is the deep soft tissues of the lower extremities, particularly the thigh and buttock. Due to its indolent growth and absence of early symptoms, ASPS often presents late, and metastatic disease may be the first manifestation. Common metastatic sites include lungs, bone, brain, and lymph nodes. Among soft tissue sarcomas, ASPS demonstrates the highest frequency of brain metastases.[1–3]

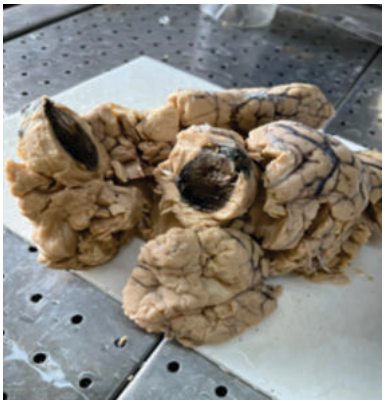

Figure 1: Gross Image of Brain Showing Metastatic Mass

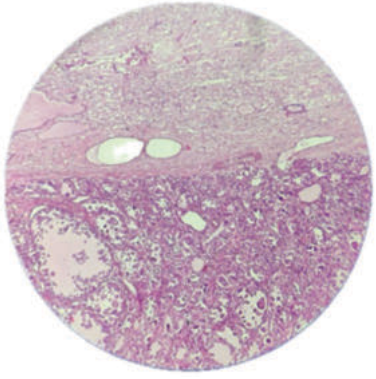

Figure 2: Histology Image Showing Normal Brain (Upper part) and Metastasis in Brain (Lower part) in High Power Field (H and E stain).

CASE STUDY

History: A patient was brought dead to Sir T. General Hospital, Bhavnagar, on 14th August 2023. Viscera were received in the autopsy section for histopathological examination following post-mortem evaluation by the forensic department.

Clinical history obtained from relatives and available medical records revealed complaints of severe headache for the past 2–3 years. The patient had previously consulted a psychiatrist, and electroencephalography (EEG) was performed, which was reported as normal. A computed tomography (CT) scan of the brain was advised but was not performed.

Past history revealed excision of a right thigh mass approximately three years prior; however, no histopathological or treatment records were available.

Gross Examination: Two well circumscribed, encapsulated, firm hemorrhagic brown masses measuring 4x3.5x5 cm and 2x1.5cm were present in left parietal lobe.

Microscopic Examination : Studied sections from both masses showed well encapsulated tumor with nesting pattern of arrangement and detachment of many tumor cells in centre and individual tumor cell have abundant, dense eosinophilic nuclei showing moderate nuclear pleomorphism and large single or two nucleoli with hobnailing appearance. Many tumor cells show clearing of cytoplasm. There are presence of extensive areas of hemorrhage. Rest of brain parenchyma shows congestion and mild oedema.

Immunohistochemistry: Immunohistochemical analysis demonstrated positivity for vimentin and focal positivity for S-100 protein and HMB-45. These findings are supportive of alveolar soft part sarcoma and assist in excluding other morphologically similar neoplasms.[1,2,4]

Discussion: Alveolar soft part sarcoma constitutes approximately 0.5–1% of all soft tissue tumors and predominantly affects young adults, with a female preponderance.[1,2] The tumor is notorious for its slow growth and lack of early symptoms, which often results in delayed diagnosis.

While lung carcinoma accounts for the highest overall incidence of brain metastases among malignancies, ASPS exhibits the highest rate of brain metastasis among soft tissue sarcomas.[1,3] This propensity explains the long-standing neurological symptoms observed in the present case.

The differential diagnosis of a metastatic brain tumor with alveolar architecture includes alveolar rhabdomyosarcoma, metastatic renal cell carcinoma, paraganglioma, melanoma, and perivascular epithelioid cell tumor (PEComa). Immunohistochemistry plays a crucial role in narrowing the diagnosis.[1,2,4]

In the present case, the history of excision of a right thigh mass three years earlier raises the possibility that the thigh lesion represented the

primary ASPS, with subsequent late metastasis to the brain. ASPS is known for delayed metastatic spread, even several years after initial tumor excision.[1]

Aggressive multimodal therapy and long-term follow-up are essential due to the high risk of distant metastasis. Prognosis depends on multiple factors including tumor size, stage at diagnosis, patient age, and presence of metastatic disease.[1,2]

CONCLUSIONS

This case highlights the aggressive metastatic potential of alveolar soft part sarcoma despite its indolent primary behavior. Brain metastasis may be the presenting or fatal manifestation, emphasizing the importance of long-term surveillance in patients with a history of deep-seated soft tissue tumors, particularly in young adults.

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