Case Report

Alveolar soft-part sarcoma presenting with multiple intracranial metastases

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A 28-year-old man presented with history of raised intracranial pressure and one episode of generalized tonic clonic seizures. Computed Tomogram revealed multiple contrast enhancing intracranial lesions. Biopsy of one of the lesions was reported as metastatic alveolar soft part sarcoma. He was advised whole brain radiotherapy.

Key Words: Brain tumor, metastases, stereotaxy

Introduction

Alveolar soft-part sarcoma, a rare tumor accounting for less than 1% of the sarcoma subtypes,[1] usually arises in the soft tissues of the extremities.[2] Its histopathogenesis is unclear but it has specific cellular characteristics. It has a propensity to metastasize to the brain late in the course of established disease. Brain metastases are unusual as the initial presentation. Only two cases of alveolar soft-part sarcoma presenting with intracranial metastases have been reported so far.[3,4] We report this case of alveolar soft-part sarcoma in which the initial presentation was multiple brain metastases.

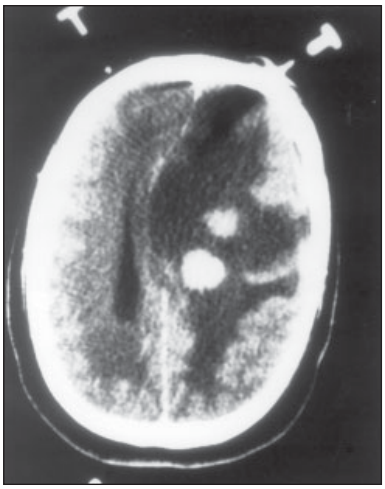
Case Report

A 28-year-old man presented with history of headache, diplopia and visual obscurations of 3 months duration and an episode of generalized tonic clonic seizures. His attention and concentration were impaired. There was bilateral papilledema, a right upper motor neuron facial weakness and impaired tandem walking. The systemic examination was normal. The chest X-ray showed multiple nodules in both the basal fields. Computed Tomogram (CT) showed multiple contrast-enhancing masses in the left frontal, left basal ganglia and right parietal regions with perilesional edema (Figures 1). Differential diagnoses of multiple abscesses and multiple intracranial metastases were considered. An ultrasound of the abdomen and a bone scan were normal. He was negative for HIV infection. He underwent a CT-guided stereotactic biopsy of the left frontal mass, which did not yield a definite histological diagnosis. Subsequently, he underwent CT-guided stereotactic right parietal craniotomy and excision of the lesion that was reported as consistent with metastatic alveolar soft-part sarcoma. The tumor brain interface was well defined. The bone and dura were normal. There was gradual worsening in his neurological status while he was in the ward. He was started on steroids and was advised whole brain radiotherapy.

The tumor had an organoid pattern with loosely cohesive polygonal cells separated into alveolar clusters by thin-walled vascular channels (Figure 2). The cytoplasm of the tumor cells contained periodic acid schiff (PAS) positive and diastase resistant crystalline and granular material. On immunohistochemistry the tumor cells were negative for epithelial membrane antigen, cytokeratin, S100, chromogranin, vimentin, actin, synaptophysin, glial fibrillary acidic protein (GFAP) and placental alkaline phosphatase.

Discussion

Christopherson et al[5] first described alveolar soft-part sarcoma as a distinct tumor of uncertain histogenesis. It is primarily a tumor of young adults with a peak age incidence



Figures 1: Contrast CT brain showing multiple ring-enhancing lesions-left frontal, left basal ganglia and right parietal regions with edema and midline shift to the right

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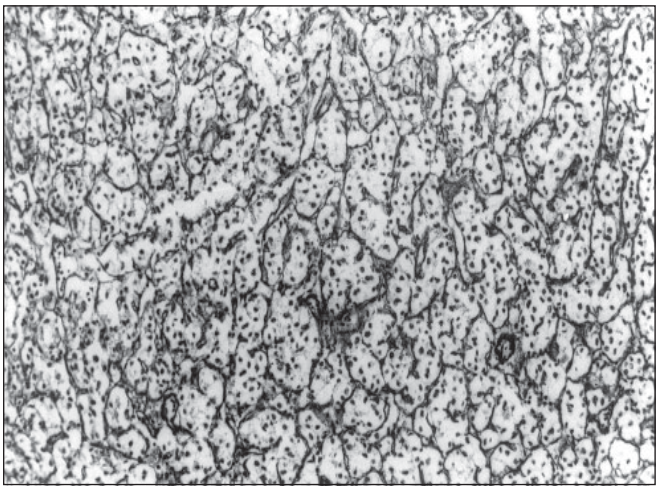

Figure 2: Photomicrograph illustrating alveolar clusters of tumor cells separated by thin-walled vascular channels. (Modified Gordon Sweet's reticulin x 90)

between 15-35 years and a female preponderance.[1] Most cases have a primary site in the lower limbs and show right-sided laterality as described by Fassbender.[6] In children the primary site is in the head and neck.[1] The histopathogenesis is uncertain but Ronald et al[7] hypothesized that alveolar soft-part sarcomas arise from displaced paraganglionic mesoderm and have a close homology with paragangliomas of the carotid body type. Most of the other studies indicate a muscle origin.[8,9] Most frequent metastases reported are lung (42%), bone (19%), brain (15%) and lymph nodes (7%).[2] In another series the incidence of brain metastases was reported as 19% and it was always noted in association with metastases to other sites.[10] Frontal lobe involvement is seen in more than 50% of cases reported.[4,11,12]

Intracranial metastases as the primary manifestation of alveolar soft-part sarcoma are very rare. Only two cases have been reported so far. Lewis[4] reported a case of a 25-year-old man with headaches who was found to have a right occipital mass lesion. The biopsy was initially reported as clear cell carcinoma. Five years later it was read as alveolar soft-part sarcoma when he presented with a thigh mass that was treated by excision. Later, Perry et al[3] reported a case of a 28-year-old man with dural-based mass lesions in the left frontal and occipital regions. The histological features of the excised frontal mass were characteristic of alveolar soft-part sarcoma. The chest X-ray of the patient showed multiple nodules in all the fields. There was no evidence of the primary site even after 18 months of follow-up.

Histologically, the tumor should be distinguished from renal cell carcinoma, granular cell tumor and paraganglioma. Histological features show alveolar clusters separated by thin-walled vascular channels. Cells are polygonal with vesicular nuclei containing a nucleolus and eosinophilic granulated cytoplasm which contains PAS positive, crystalline to granular material. In children the cells are compactly arranged.[1] Recent cytogenetic analysis has revealed chromosome rearrangements at 17q25 and Xp11.[2,13]

Previous case reports show that surgically treated cases have a favorable outcome.[4,11] Bindal et al[12] recommend surgical excision of the intracranial metastases in patients who are not terminally ill and do not consider involvement of lungs as a contraindication for surgery and they found that the five-year survival in these patients is better than metastases from other histologies. Radiotherapy is recommended after surgical excision. It is reported that metastatic alveolar soft-part sarcoma is resistant to conventional doxorubicin-based chemotherapy.[10] As our patient's condition was rapidly deteriorating he was referred for whole brain radiotherapy.

In conclusion, alveolar soft-part sarcoma accounts for a significant proportion of sarcomas that metastasize to the brain and need to be kept in mind when one encounters a case with typical histology. Early surgical excision followed by radiotherapy improves survival.

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