Bleeding jejunal metastases arising from primary alveolar soft part sarcoma of thigh

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ABSTRACT

Alveolar soft part sarcoma (ASPS) is a rare tumor (0.5–1% of sarcomas) of unknown etiology with a highly characteristic morphology. It tends to occur more often in the younger age group with a predilection for the female sex. Prognosis is generally poor and it often presents with late metastases. Here, we report the case of a 48-year-old female, who presented to the emergency department with a complaint of giddiness and melena. She was a known case of sarcoma ASPS left thigh with pulmonary secondaries. On evaluation, a bleeding tumor in the upper jejunum was found on upper GI endoscopy. As the patient's hemodynamics were unstable, she had to be operated without further imaging. Post-operative period was uneventful.

Keywords: *Alveolar soft part sarcoma, Bleeding tumor, Jejunum, Metastases*

Alveolar soft part sarcoma (ASPS) is a rare tumor (0.5–1% of sarcomas) of unknown etiology with a highly characteristic morphology. It was first described and named by Christopherson and Stewart, in 1952 [1,2]. It has an unusual biologic behavior with a relatively indolent course. In general, it has a poor prognosis due to widespread metastases (68%). It most often originates in the thigh or buttocks in adolescents and young adults, especially females, with a proclivity for metastases to the lung, bone, and brain [3]. Hematogenous metastasis to the gastrointestinal (GI) tract from ASPS is an extremely rare occurrence with only a few cases reported in the literature [4-8].

We report the case of a 48-year-old female, who was a known case of ASPS of the left thigh with pulmonary secondaries, presented to the emergency department with a complaint of giddiness and melena and later on found a bleeding tumor in the upper jejunum.

CASE REPORT

A 48-year-old female presented to the emergency department with a chief complaint of giddiness and melena for the past 1 day. She was a known case of primary ASPS left thigh with pulmonary secondaries. She gave a history of surgery for the tumor in the anterior aspect of left thigh in 1992. Postoperatively, she received adjuvant radiotherapy and chemotherapy (no details regarding dosage and frequency available). She developed pulmonary metastases after 5 years, for which she received palliative chemotherapy (3 weekly Injection epirubicin [D1 and D2] and Injection ifosfamide [D1, D2, and D3] with mesna chemotherapy for 6 cycles). She was symptom free for the next 10 years, after which she again received chemotherapy for worsening of pulmonary symptoms (Injection gemcitabine based). The patient underwent palliative sleeve right lobectomy 7 years later (2014) after the last chemotherapy. The patient was not willing for further chemotherapy which was advised.

Now, she presented to our department with the above complaints. On clinical evaluation, the patient was hemodynamically unstable with a pulse rate of 140/min and blood pressure of 80/40 mmHg. Rest of the vitals were normal. Routine blood investigations were performed, which revealed severe anemia (Hb–4.5 g/dl; normal –12-14 g/dl) and serum creatinine levels of 1.6 mg/dl (0.9–1.2 mg/dl). Arterial blood gas analysis revealed metabolic acidosis picture with a pH value of 7.23, lactate value of 4.6 mmol/l (0.5–1 mmol/l), and bicarbonate levels of 16 mmol/l (22–24 mmol/l).

Blood transfusion and inotropes were started and upper GI endoscopy was attempted after securing the airway. The procedure was abandoned as the view was obscured by blood clots. Once the condition was stabilized with blood transfusions and inotropes, repeat endoscopy was done using push enteroscopy, which showed actively bleeding tumor about 50 cm from duodenojejunal flexure not amenable to endoscopic measures (Fig. 1). No imaging was possible due to a hemodynamically unstable condition. She was taken up for emergency laparotomy after four blood transfusions, and a palliative resection of the jejunal tumor was done (Fig. 2). Postoperatively, two more blood transfusions were done. The patient was weaned off from the ventilator after 72 h.

Histopathology report showed that the tumor cells were polygonal with abundant granular eosinophilic cytoplasm and

arranged in nests of tumor cells, separated by a thin fibrous septae. Central degeneration and loss of cohesion of cells in the center of the nests create the characteristic "pseudoalveolar" pattern (Fig. 3). Immunostain positivity was seen only for desmin and all other immunostains were negative (Fig. 4). At present, the patient is receiving palliative chemotherapy (eribulin based) and is doing well.

DISCUSSION

ASPS is a rare tumor of unknown etiology with highly characteristic morphology. It is characterized by uniform, organoid nests of polygonal tumor cells, separated by fibrovascular septa, and delicate capillary-sized vascular channels with prominent cellular dyscohesion, leading to the distinctive pseudoalveolar pattern [1]. ASPS has a non-reciprocal chromosomal translocation der (17) t (X; 17) (p11; q25) with the corresponding oncogenic fusion gene, alveolar soft part locus - transcription factor E 3 [9].

There is a predilection for young females, especially during the first 2 decades of life with a female-to-male ratio of 2:1 [1], and it most commonly involves the deeper soft tissue of the extremities, trunk, head, neck, and retroperitoneum. The most important prognostic factors are the age at diagnosis (younger patients have a better prognosis), size of the tumor (larger tumors have a worse prognosis), and the presence of metastatic disease at presentation [9,10]. A characteristic finding on electron microscopy is the presence of periodic acid-Schiff-positive, diastase-resistant crystalline structures, which may be rhomboid, rod-like or spiked, in individual, and sheaf-like or stacked configurations. Immunohistochemistry can be helpful and is usually reactive for vimentin, muscle-specific actin, desmin, cytoplasmic MyoD1 and usually non-reactive for pan-cytokeratin, synaptophysin, chromogranin, and myogenin [11].

The differential diagnosis of ASPS to be considered is rather broad and comprises neoplasms that may show organoid patterns of growth and cells with abundant eosinophilic cytoplasm which includes renal cell carcinomas, adrenal cortical carcinomas and hepatocellular carcinomas, malignant melanoma, and paraganglioma. Immunohistochemistry plays a major role in differentiating these entities (strong cytokeratin expression, expression of markers such as renal cell carcinoma antigen in renal cell carcinoma, HepPar1 in hepatocellular carcinoma, S100 protein, human melanoma black 45, and melan A in melanoma). Paragangliomas show strong expression of chromogranin A and synaptophysin, unlike ASPS. Alveolar rhabdomyosarcoma

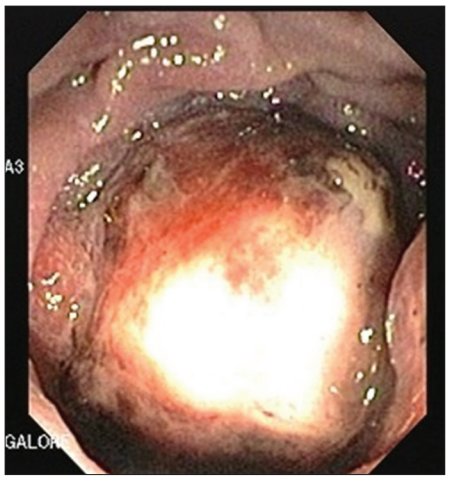

Figure 1: Upper gastrointestinal endoscopy showing intraluminal lobulated soft tissue mass in jejunum 50 cm from duodenojejunal flexure

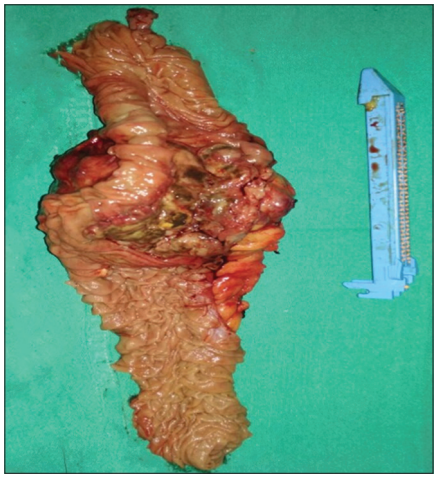

Figure 2: Resected segment of jejunum with tumor

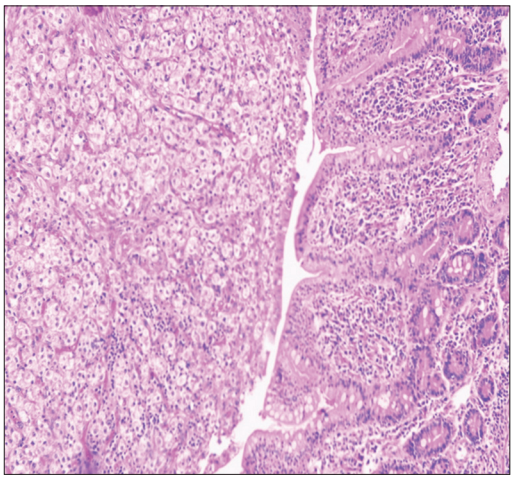

Figure 3: Histopathology of the resected tumor

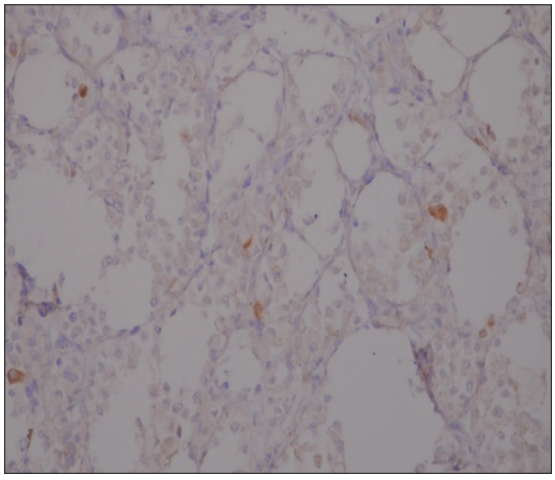

Figure 4: Immunostain: Focal desmin positivity

strongly expresses desmin and myogenin nuclear regulatory proteins need to be considered in the differential diagnosis [12].

The presence of metastases does not preclude a long survival time [3]. Surgical resection of the metastases appears to increase the median survival and should be considered [13]. There are several reports in the literature of long-term survivors with metastatic ASPS. One case report describes a patient who developed the primary chest wall lesion in 1960, pulmonary metastases in 1981, and brain and renal metastases in 1992 [14]. This report and others [14-16] suggest that in carefully selected patients, aggressive and repeated treatment of the metastatic disease may influence long-term survival and maintenance of good performance status in patients [7]. The patient described in our report has 27 years of survival from the time of her initial diagnosis to this most recent episode. The GI tract is uncommonly affected by primary or metastatic ASPS with only a few cases being reported in the literature [4-8,17].

The first case of GI metastases was reported by Sueyoshi, in 1996, affecting jejunum with GI bleeding associated [5]. In 2001, Sabel *et al*. described a case in the small bowel, causing polyposis and intussusception in a 42-year-old male with the previous history of primary ASPS of thigh metastatic to lung and brain [7]. The patient was alive at 15 years after the primary diagnosis till the latest episode of intestinal metastases. In 2003, Zilber *et al*. found the first case of colic metastases in a 43-year-old woman with a primary leg tumor more than 5 years before and multiple lungs and brain metastases. She was found to have metastases in the cecum, who presented with anemia and was treated by laparoscopic right colectomy [8]. In few of the cases reported [7,8,18] and also in the present case, the sequence of the metastases observed was lung followed by brain and GI tract. Hence, one needs to be cautious during the follow-up period to focus on these areas to look for metastases.

All previously reported cases share similar clinical features, but our case had an acute presentation with hemorrhagic shock requiring immediate emergency surgery. As it might metastasize to the GI tract and induce substantial bleeding, a high index of suspicion must be maintained for the patients with a history of alveolar sarcoma presenting with complaints of hematemesis or melena and a prompt endoscopy, abdominal computed tomography scanning followed by immediate laparotomy if needed should be carried out in such patients.

CONCLUSION

Histology by itself predicts the prognosis of the disease. Primary ASPS is one such disease, which has an indolent course and is amenable to multimodality treatment. The aggressive form of treatment needs to be considered even in a metastatic setting when such disease is encountered.

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