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DUODENAL METASTASIS OF ALVEOLAR SOFT PART SARCOMA

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Aveolar soft part sarcoma is a rare tumor responsible for about 1% of all soft tissue sarcomas, affecting mostly adolescents and young adults. ASPS has curious patterns of metastatic spread, with seldom lymph node involvement. Lung, bone and brain are the most common metastatic places. Small bowel metastasis are infrequent, having found reported only one case of duodenal metastasis with polypous appearance. We describe a case of duodenal metastasis presenting as abdominal mass five years after initial diagnosis of alveolar soft part sarcoma.

Key-word: Sarcoma.

Introduction

Alveolar soft part sarcoma (ASPS) is an uncommon soft tissue neoplasm affecting mostly adolescents and young adults with a slight female predominance. The tumor grows slowly, but is well-vascularized with a tendency to perform vascular invasion and hematogenous distant metastases. The most common metastatic places are lung, bone and brain. Involvement of gastrointestinal tract is very rare; four cases have been described in literature and only one duodenal metastasis has been reported. Duodenal metastases are very uncommon. We describe a case of duodenal metastasis in appearance of a mass five years after initial diagnosis of alveolar soft part sarcoma.

Case report

A 27-year-old woman with a 3 day history of palpitations was admitted to our hospital. She complained of effort related nausea, fatigue, headache, muscular weakness asthenia and one month weight loss. She had history of alveolar soft part sarcoma on her right leg five years ago, treated with radiotherapy. Two years ago she underwent lung metastasis resection, with no evidence of other metastases at the time. Physical examination revealed pale skin and hepatosplenomegalia. Blood sample analysis showed haemoglobin of 5,5 g/dL and hematocrit of 16,8%: anaemia. A multidetector CT abdominal scan without contrast (allergy antecedents) demonstrated a round homogeneous hypodense alteration (mass) of the duodenum with pancreas displacement (Fig. 1). Additional imaging was necessary. The MRI study (Fig. 2, 3) was performed with a Signa Excite XI Twin Speed 1.5T system (GE Healthcare, Milwaukee, WI, USA) using FRFSE T2-weighted, FRSPGR (phase and out of phase) and LAVA sequences. MRI showed a 47 mm mass delimited by serosa at the second duodenal portion. The mass was slightly hyperintense with strong central hyperintense area suggesting necrosis and early heterogeneous enhancement in T2-weigthed sequence. The necrotic area presented luminal communication to duodenum. No evidence of pancreas head infiltration was

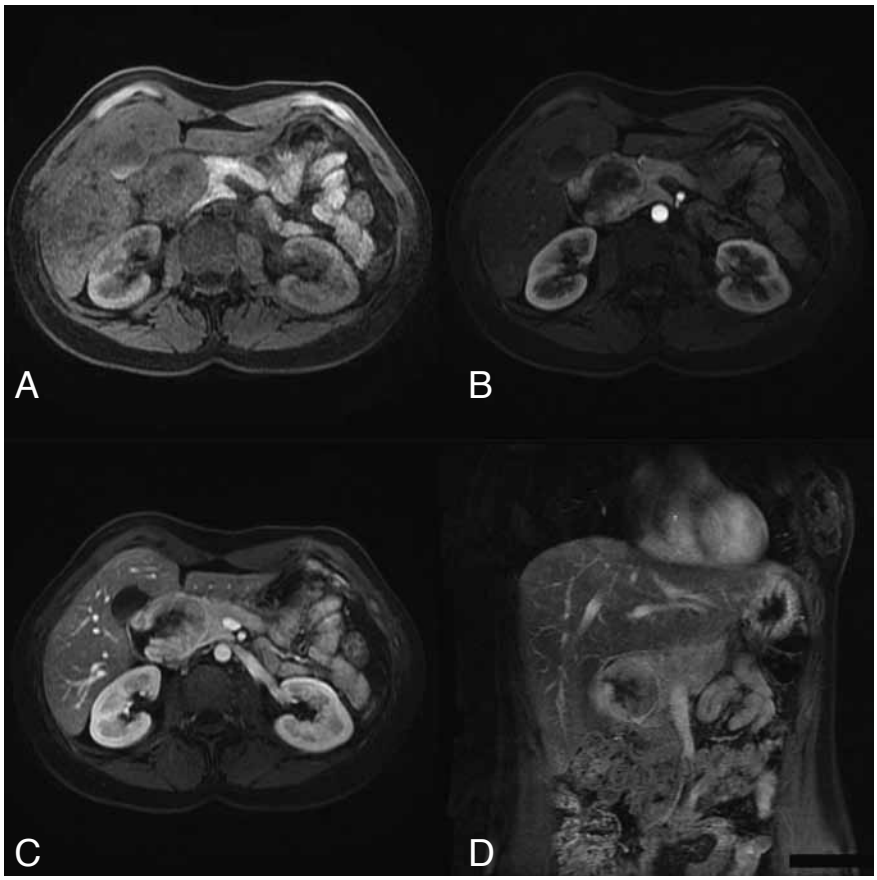

Fig. 1. — Axial (A) T1-weighted MRI image after contrast shows an isointense mass displacing the pancreatic head. The axial MRI image in arterial phase (B) demonstrates early peripheral enhancement in the pancreatic site of the mass with a central hypointense area. The arrow reveals the normal duodenal external wall. Images C, axial and D, coronal, shows the portal phase with progressive enhancement of the periduodenal well-delineated rounded mass with peripheral enhanced ring.

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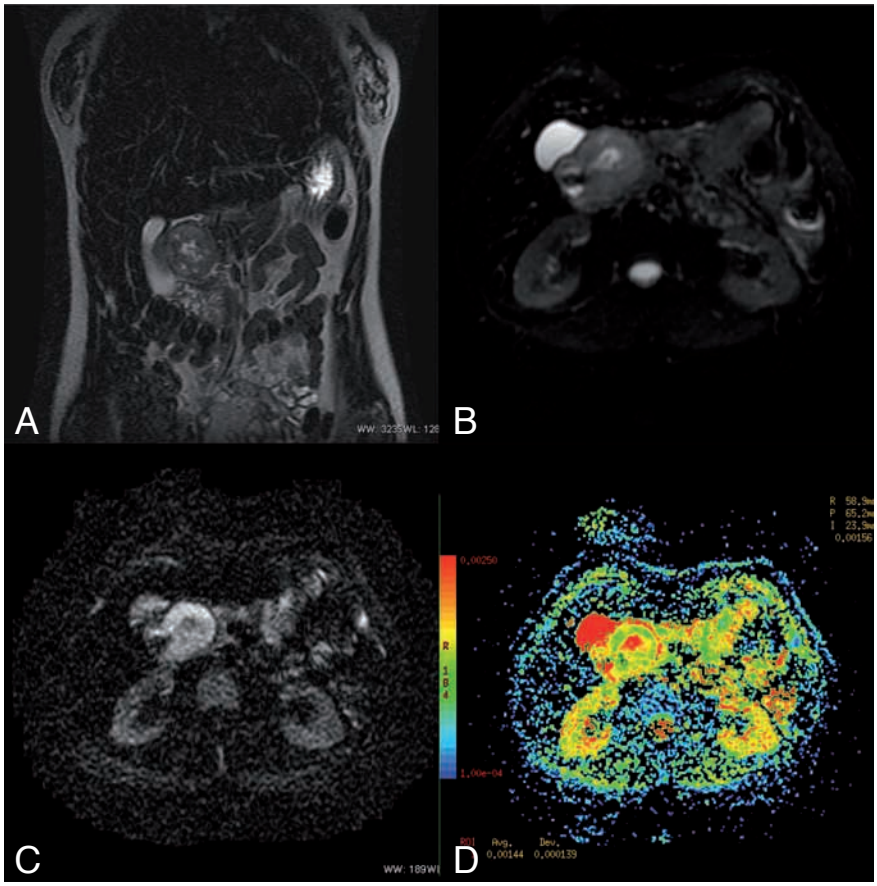

Fig. 2. — The coronal T2-weighed image (A) reveals a slight intense mass with central hyperintense area suggesting a necrotic area. Image B is a proton density MRI image with B-factor 0. The proton density MRI image with B-factor 1000 (C) shows high signal at the mass suggesting of a high restriction mass. Image D demonstrates the apparent diffusion coefficient (ADC) of 0.00144, which is a low coefficient.

found. Surgical treatment consisted in laparoscopic Wipple duodenopancreatectomy with removal of the duodenal mass. Histopathological examination demonstrated an ASPS metastasis. The patient underwent a control chest-abdominal CT 2 months later that showed no evidence of tumor, adenopathy nor metastasis.

Discussion

Alveolar soft part sarcoma (ASPS) was first described by Christopherson et al. in 1952 (1). It is a rare type of soft tissue malignant tumor which accounts for about 1% of all soft tissue sarcomas (2). The disease affects mostly adolescents and young adults, with a lower age at diagnosis compared to other forms of soft tissue sarcoma. The peak incidence is described between 15 and 35 years old (3). There is a female predilection, especially during the first two decades of life (4). In adults it most commonly involves the muscle and deep soft tissue of extremities, trunk, head, neck and retroperitoneum. Head and neck locations are more commonly affected in children and adolescents (5, 6).

ASPS has unusual patterns of metastatic spread (7). Metastasis occurs in about 68% of cases and is primarily haematogenous, with rarely lymph node association (8). The most common metastatic places are lung, bone and brain (9).

The gastrointestinal tract is uncommonly affected by ASPS metastases. Only four cases of ASPS intestinal metastases has been reported in literature so far, the first by Sueyoshi in 1996, affecting jejunum with gastrointestinal bleeding associated (10). In 2001, Sabel et al. described a case in small bowel, causing polyposis and intussusception in a 42-year-old male with previous history of ASPS metastatic to lung and brain (11). Zilber et al. in 2003 found the first case of colic metastases in a 43-year-old woman with a leg primary tumor more than 15 years before and multiple lung and brain metastases. She was found to have caecal metastases, revealed by anaemia, and was treated by laparoscopic right colectomy (12). In 2009, Banihani et al. published a case about a 38-year-old man with a huge abdominal mass infiltrating the omentum. Pathological diagnosis was ASPS. He had metastases in both lungs and the right atrium. Afterwards multiple sessile polyps also appeared in stomach and duodenum with diagnostic biopsy of ASPS. Finally the patient developed brain metastases and died (13).

Primary gastrointestinal ASPS is extremely rare. Only one case has been reported in 2000 by Yaziji et al., a primary ASPS of the stomach in a 54-year-old Italian woman without evidence of primary neoplasm elsewhere ten years following the initial diagnosis (14).

In metastatic tumors, small bowel involvement is uncommon and has been described in only 2% of autopsy cases. Secondary tumors involving the duodenum can arise from peritoneal dissemination, direct spread from an intra-abdominal malignancy, hematogenous and lymphatic spread (15). Common metastatic malignancies known to involve the small intestines are melanomas, lung cancer (16), cervix carcinoma, renal cell carcinoma, thyroid carcinoma, hepatocellular carcinoma and Merkel cell carcinoma (17). The incidence increases with age and males are more commonly affected. Metastatic lesions of the duodenum mostly locate in the periampullar region, followed by the duodenal bulb. Patients present with abdominal pain, nausea, vomiting and gastrointestinal bleeding (18).

The microscopic picture of ASPS is uniform and characterized by a pseudoalveolar pattern with nests of tumor cells separated by sinusoidal vascular channels. The cells have vesicular nucleoli and eosinophilic cytoplasm (3).

Magnetic resonance imaging is the best technique for characterization of ASPS. Common MRI findings are high-signal-intensity on T1 and T2-weighted images and multiple intra- and extra-tumoral signal voids. The high-signal-intensity areas of the tumors on T1-weighted sequence can be attributed to slow flowing blood in or around the tumor (19).

Presence of metastases at the time of diagnosis carries a poorer prognosis (median survival time of 3 years), while early metastases do

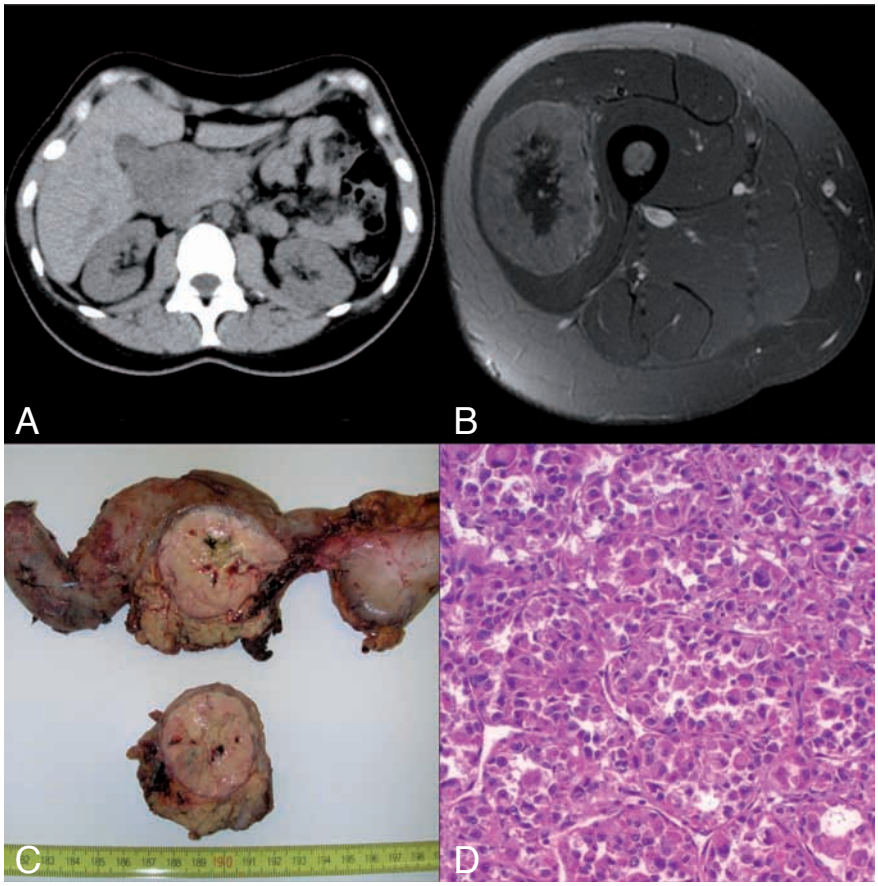

Fig. 3. — An axial CT image without contrast (A) shows a round mass in the pancreatic area. B is the MRI image (B) of the initial ASPS tumor in the leg. The mass shows peripheral enhancement and a central hypodense area suggesting necrosis. The macroscopic image (C) shows macroscopic piece of cephalic duodenopancreatectomy with a 6 cm circumscribed mass within the duodenopancreatic space. Both the 4 cm duodenal ulceration and the area of central necrosis suggest malignancy. Pancreas structure is displaced and the limits of surgical resection are clear. The microscopic image (D) reveals the malignant tumor with a clear alveolar pattern defined by nests of peculiar large sized cells, separated by a delicate slightly vascularized connective tissue. Large polygonal cells with a wide cytoplasm, large nuclei and evident macronucleoli provide polymorphic appearance.

not preclude a long survival time (8). The treatment of choice of ASPS primary and metastatic tumors remains on surgical resection. The benefit of adjuvant chemotherapy and/or radiotherapy has been doubtful (20).

In conclusion, ASPS is a rare type of sarcoma that affects mostly the lower limbs. This tumor does rarely metastasize to the gastrointestinal tract. We report the second case of duodenal metastasis, which developed five years after the initial diagnosis.

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