DUODENAL METASTASIS OF ALVEOLAR SOFT PART SARCOMA

I. Willekens1, C. Paradisi3, L. Sarria3, A. Puertas4, J. Pac4, E. Mayayo4

Aveolar soft part sarcoma (ASPS) is a rare tumor with a predilection for adolescents and young adults. The tumor typically 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 rare; only one duodenal metastasis has been reported. We describe a case of duodenal metastasis in a 27-year-old woman five years after initial diagnosis of alveolar soft part sarcoma.

Key-word: Sarcoma.

Introduction

Alveolar soft part sarcoma (ASPS) 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 as an abdominal 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, 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 hepatosplenomegaly. 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 hypo dense 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-weighted sequence. The necrotic area presented luminal communication to duodenum. No evidence of necrotic area presented luminal communication to duodenum. No evidence of necrotic area presented luminal communication to duodenum. No evidence of necrotic area presented luminal communication to duodenum.

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, show the portal phase with progressive enhancement of the periduodenal well-delineated rounded mass with peripheral enhanced ring.

From: 1. In vivo Cellular and Molecular Imaging, ICMI, Vrije Universiteit Brussel, Brussels, Belgium. 2. Department of Radiology, UZ Brussels, Brussels, Belgium. 3. Department of Radiology, 4. Department of Pathology, Hospital Universitario Miguel Servet, Zaragoza, Spain.

Address for correspondence: Dr I. Willekens, M.D., Department of Radiology, UZ Brussels, Laarbeeklaan 101/103, 1090 Brussels, Belgium.

E-mail: Inneke.willekens@gmail.com
com m only involves the muscle and decades of life (4). In adults it most
tion, especially during the first two
described between 15 and 35 years
sue sarcoma. The peak incidence is
com pared to other forms of soft tis-
adults, with a lower age at diagnosis
accounts for about 1% of all soft tis-
as first described by Christopherson
metastasis. The patient underwent a
control chest-abdominal CT
months later that showed no evi-
dence 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 Suyeshi in 1996, affecting jejenum 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 2008, Baniani 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 intestine 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 nuclei 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. 2. — The coronal T2-weighted 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) show s 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.
References


