Pediatric Primary Splenic Angiosarcoma: A very rare disease

Angiosarcome splénique chez l'enfant: Une maladie exceptionnelle

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RÉSUMÉ

L'angiosarcome splénique primaire est une tumeur rare et agressive. Diagnostiquer principalement chez les adultes de plus de 40 ans. Il ya eu moins de dix cas pédiatriques rapportés. Une fille de 13 ans s'est présentée aux urgences pour douleur du quadrant supérieur gauche avec fatigue. Les résultats de laboratoire ont révélé une légère anémie (11,3 g / dL) avec des plaquettes normales (166. 109 / L). Une formation hétérogène avec un grand axe de 5 cm a été identifiée sur le scanner de l'abdomen. Une Splénectomie par laparotomie a été réalisée, et un angiosarcome splénique primaire a été découvert. Sept mois plus tard, un scanner de control n'a pas montré une maladie progressive, en particulier, pas de métastases au foie. Elle est actuellement en vie avec des signes de maladie à sept mois, mais sans progression. L'Angiosarcome splénique primaire est Presque universellement fatale, malgré le traitement. La meilleure chance de survie est le diagnostic précoce et la splénectomie avant la rupture de la rate.

Mots-clés

Hémangiosarcome, Cancer de la rate

SUMMARY

The primary splenic angiosarcoma is a rare and aggressive tumor. Diagnose primarily in adults over 40 years. There were less than ten pediatric cases reported. A 13-year-old presented to the emergency with left upper quadrant pain. Laboratory results revealed anemia (11.3 g / dL) with normal platelets (166. 109 / L). A heterogeneous formation with a major axis of 5 cm has been identified on the scan of the abdomen. Splenectomy by a laparotomy was performed, and angiosarcoma Primary splenic was discovered. Seven months later, a scanner control showed no progressive disease, in particular, no liver metastases. She is currently alive without signs of progression. The primary splenic angiosarcoma is almost universally fatal, despite treatment. The best chance of survival is early diagnosis and splenectomy before the breaking of the spleen.

Key-words

Hemangiosarcoma, Splenic cancer

Primary splenic angiosarcomas are among the rarest types of neoplasm, with an estimated annual incidence of 0.14 to 0.25 cases per million persons (1).They are aggressive malignant neoplasms arising from splenic sinusoidal vascular endothelium. Upper left abdominal pain, weight loss, fatigue, generalized weakness and fever are common symptoms (5). The morphologic spectrum of this disease is highly variable being similar in some aspects to vascular tumors such as hemangiomas, malignant non-vascular tumors like lymphangiomas and secondary metastatic cancers, causing diagnostic difficulty. Splenectomy is thought to be the only intervention that may result in long-term, disease-free survival. Almost all patients die within 12 months of diagnosis regardless of treatment (4). They have proven to be highly refractory to adjuvant radiation and chemotherapy. Very few reviews are available to date in the literature. A case report of this rare clinical entity and complete review of the current literature are provided.

CASE REPORT

A 13-year-old girl was admitted with left upper quadrant pain and fatigue. There was no history of chemical agent exposure but there was a history of mild trauma in left upper quadrant 2 months ago. Physical examination revealed sensibility in left upper quadrant. Anemia was present (HB: 11.3 g/dl). An abdominal ultrasonography (US) revealed a mass of 5cm major axis with hypoechogene and heterogeneous echogenicity in the spleen (Fig 1A). The liver was normal. Subsequent CT examination of the whole abdomen confirmed the sonographic findings. The 5cm mass of the spleen was hypodense on CT scans. Calcification was not present (Fig 1B). The splenic parenchyma was heterogeneous. The patient underwent laparotomic splenectomy.

Figure 1A : Abdominal ultrasonography: Hypoechogene mass in the spleen

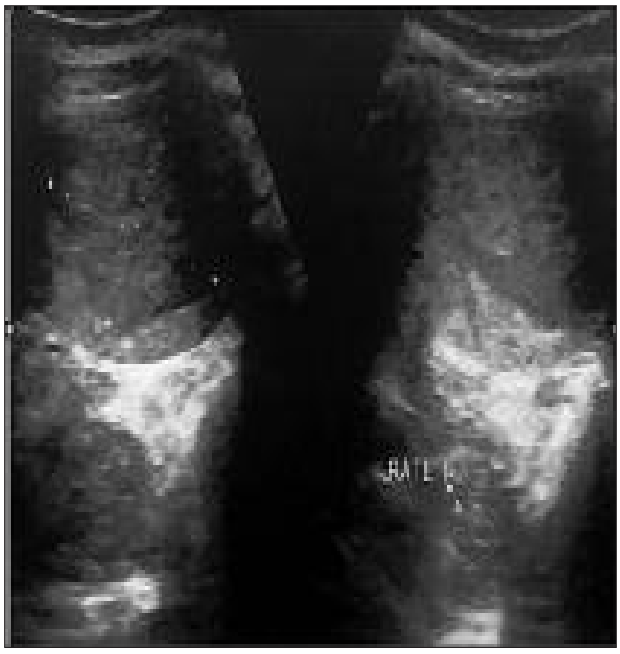


Figure 1B : CT: Hypodense mass in the spleen

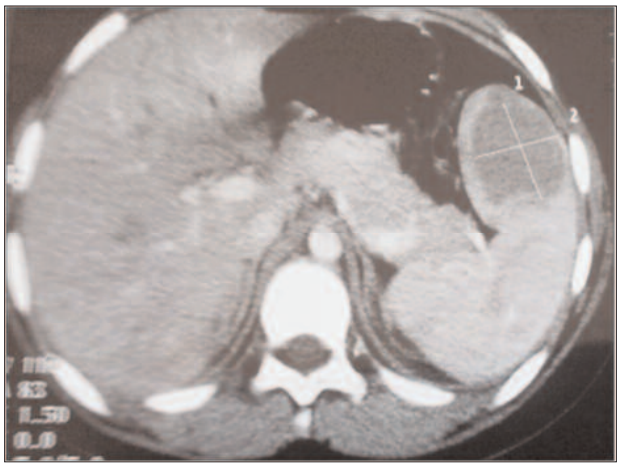
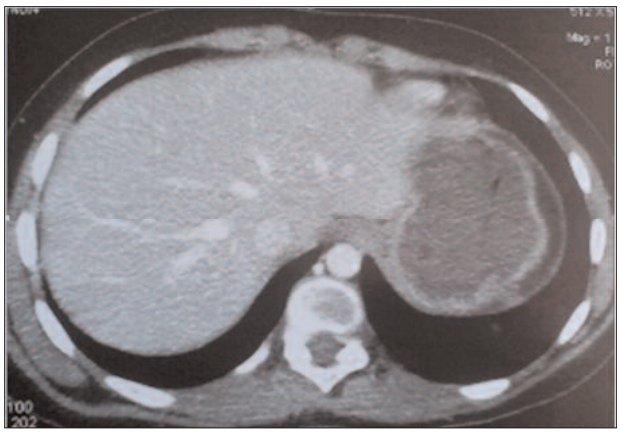


Figure 1C : CT after seven months: No liver metastasis



The specimen measured 5*4 cm. Histopathologic examination disclosed primary angiosarcoma. The patient hasn't received any course of chimiotherapy. Follow-up CT imaging at seven months after surgery revealed no metastatic lesions in the liver (Figure 1C). The patient is still alive without complications.

DISCUSSION

Angiosarcomas are rare soft-tissue sarcomas of endothelial cell origin that have a poor prognosis (6). The mean age at presentation is 59 years with a range between 14 months and 89 years (1). There have been eight reported pediatric cases (7). Left upper abdominal pain is the most common symptom. It occurs in 75% to 83% of patients with splenic angiosarcoma (1). Splenomegaly is the most common finding, in 68% of cases, at physical examination (8). The major complication is splenic rupture, which leads to fatal hemoperitoneum in up to 30% (9), can also be a presenting finding. Anemia is the most common laboratory abnormality in 75% to 81% of cases (10). Patients with splenic angiosarcoma often have spleens weighing more than 1000g (11).In histology, the tumor consists of disorganized anastomosing

vascular channels lined by large, atypical endothelial cells with significant irregular, hyperchromatic nuclei. The pathogenesis of primary splenic angiosarcoma is still unknown. Some authors reported causes of primary splenic angiosarcoma to exposure to ionizing radiation, chemotherapy or to some chemical agents, such as thorium dioxide, vinyl chloride, and arsenic (7). Moreover, some authors claim that these tumors develop from preexisting benign counterparts, such as hemangioma or hemangioendotheliomas (7). No such preexisting risk factors were present in our patient. The radiologic features of splenic angiosarcomas are variable and nonspecific. On ultrasound, a complex mass with heterogeneous echotexture is the most common finding. CT may reveal a heterogeneously enhancing splenic mass with areas of necrosis, the tumors may exhibit peripheral or heterogeneous contrast enhancement, on contrast-enhanced CT scans (3, 11). Primary splenic angiosarcoma has a high rate of metastatic disease, ranging from 69-100% (12). The median of survival is five months irrespective of treatment (7). However, early diagnosis with splenectomy has a favorable survival rate. Hsu et al. (13) reported that a patient in their study remained disease-free 162 months after splenectomy. This patient is the longest survivor of splenic angiosarcoma in the literature. Biopsy is contraindicated in splenic angiosarcoma because of high risk of rupture. Therefore, a definitive diagnosis can only be made by histopathologic examinations after splenectomy.

CONCLUSION

Primary angiosarcoma of the spleen is rare, aggressive and associated with a very poor prognosis. Its clinical and radiologic diagnoses are challenging. Chemotherapy has historically been unsuccessful in improving its outcome. The best chance for survival is early diagnosis and prompt splenectomy prior to splenic rupture. So, although rare, the possibility of angiosarcoma should be kept in mind in assessing splenic tumors.

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