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Pancreatic pleomorphic rhabdomyosarcoma

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ABSTRACT

INTRODUCTION: Rhabdomyosarcoma (RMS) is a primary malignancy that arises from the embryonic mesenchyme with the potential to differentiate into skeletal muscle. RMS of the biliary tree is extremely rare. We report a case of an undifferentiated pleomorphic RMS involving the liver and pancreas.

PRESENTATION OF CASE: A 62 year old Caucasian woman with rapidly growing abdominal mass and a history of endometrial adenocarcinoma underwent laparotomy due to compression symptoms and concerns of malignancy. A large mass arising from the pancreas and extending into the liver was identified and resected with a distal pancreatectomy associated with a left lateral liver segmentectomy. A diagnosis of pleomorphic RMS was made from the pathology specimen. Chemotherapy and radiotherapy were also performed. Unfortunately the patient died 2 years following treatment due to recurrence of the disease.

DISCUSSION: P-RMS in the biliary tree is extremely rare (0.5%) and mostly seen in infants and children. Preoperative diagnosis is challenging since the symptoms are nonspecific. Preoperative imaging rarely contributes to the final diagnosis. The only possible treatment for adult RMS is surgical resection of the tumor followed by chemotherapy and radiotherapy. Long-term prognosis of P-RMS reported (predominantly of limbs) is poor. To our knowledge, no previous cases of RMS originating from the pancreas have been reported.

CONCLUSION: However RMS is an extremely rare tumor in adults, it should be included in the differential diagnosis of patients with atypical pancreatic and liver lesions.

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1. Introduction

Rhabdomyosarcoma (RMS) is a primary malignancy that arises from the embryonic mesenchyme with the potential to differentiate into skeletal muscle [1]. While it constitutes approximately 50% of all soft tissue sarcomas in children, it is rare in adults accounting for approximately 3% of all soft tissue sarcomas usually localized in skeletal muscle [2–4]. RMS was described for the first time by Webber in 1854 and later classified into four histopathological subtypes: embryonal, botryoid, alveolar, and pleomorphic [1,4,5].

RMS of the biliary tree is extremely rare (approx. 0.5%) and encountered mostly in infants and children [6] and it has been described in four patients in relation to the gallbladder [7]. We report a case of an undifferentiated pleomorphic RMS involving the pancreas.

2. Case presentation

A 62 year old Caucasian woman referred to our center for rapidly growing epigastric mass. Her past medical history was significant for endometrial Cancer (Papillary Serous endometrial carcinoma, Grade 1, FIGO IIb) two years prior treated by surgical resection and radiotherapy. During the evaluation, an abdominal CT scan revealed a large abdominal mass (11.5 × 9.4 × 12.1 cm) with irregular peripheral enhancement that appears to arise from left lobe of the liver (Fig. 1A). It appeared compressing the portal vein and the stomach. Ultrasound guided biopsy was performed and revealed necrosis with rare degenerate atypical cells. Indications for surgical intervention were concern of malignancy and compression symptoms. At surgery the mass had a capsular structure with numerous vascular adhesions to the surrounding tissues (Fig. 1B). Left lateral segmentectomy and distal pancreatectomy were performed to remove the mass since macroscopically it appeared to invade both organs (Fig. 1C and D). The patient recovered uneventfully. At pathology the lesion measured (19 × 16.5 × 5.5 cm) with extensive hemorrhage and necrosis. A fine fibrous septum separated the tumor tissue from the adjacent liver and pancreas parenchyma with an interruption only in the pancreas (Stage III, Group I according to the International RMS Study Group; IRSG). Immunohistochemical

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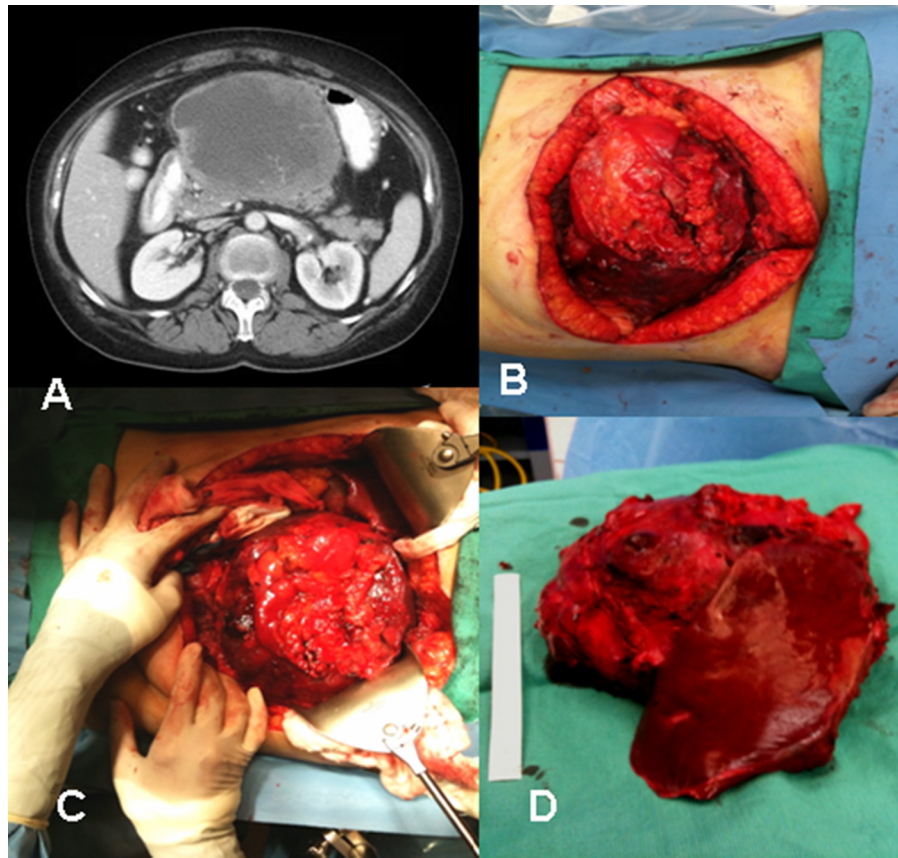


Fig. 1. (A) CT scan showing an 11.5 × 9.4 × 12.1 mass with irregular peripheral enhancement. (B and C): Epigastric mass on exploratory Laparotomy. (D) Mass resected enblock with tail of the pancreas and the left lateral segment of the liver.

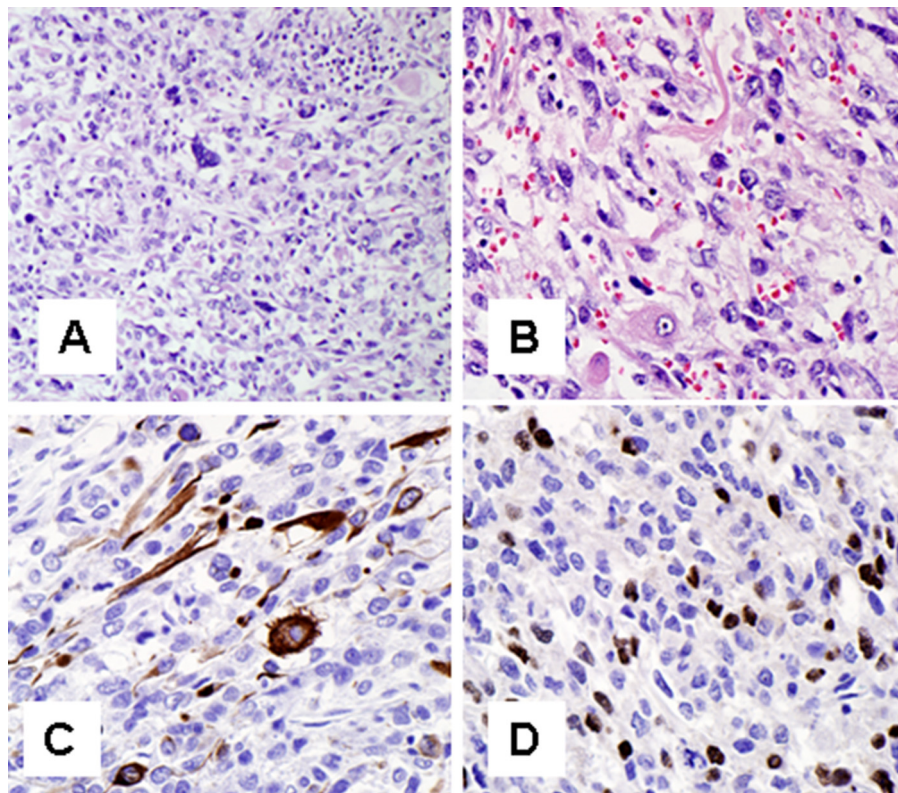


Fig. 2. (A) Pleomorphic rhabdomyosarcoma with haphazardly spindle cells and scattered cells with large eosinophilic cytoplasm and pleomorphic nuclei. (B) Elongated rhabdomyoblasts with distinct cross striation and large eosinophilic stringy cytoplasm. (C) Neoplastic cells show reactivity for Desmin. (D) Neoplastic cells stained for Myogenin.

staining was positive for vimentin, desmin, myogen and SMA and negative for cytokeratin AE1/AE3, EMA and S100 (Fig. 2). A definitive diagnosis of pleomorphic rhabdomyosarcoma (P-RMS) was made. A chemotherapy regimen with cyclophosphamide, dactinomycin and vincristin started and continued for 4 month. Along with chemotherapy radiotherapy of the resection area was performed. Patient was doing well for almost two years but she developed symptoms of compression on the stomach such as fullness, nausea and vomiting. Following further workup interval development of a soft tissue lesion along the ventral surface of the pancreatic head/neck, which was invading the adjacent gastric wall were seen. Findings were consistent with locoregional recurrence that was confirmed by tissue biopsy and was unresectable at this time, the patient died 2 month later due to extent of the disease.

3. Discussion

In contrast to what has been described mostly for extremities of adult males, P-RMS of the liver and pancreas represents an extraordinary rare entity. P-RMS in the biliary tree is extremely rare (0.5%) and mostly seen in infants and children [6]. There are only 2 cases of P-RMS of the liver have been reported in the literature in adults [8,9]. Ali et al. also reported a case of biliary RMS mimicking a choledochal cyst [10]. We have no knowledge of any cases of P-RMS arising from the pancreas in adults.

RMS arises from embryonic mesenchyme with the potential to differentiate into skeletal muscle. Approximately half of all soft tissue sarcomas in children are RMS while in adults are only 3% [4]. The origin of the tumor in our case appears to be pancreatic since the interruption of the separating fibrous tissue was seen only in the pancreatic tissue and not in the liver.

Preoperative diagnosis is challenging since the symptoms are unspecific with abdominal discomfort, weight loss and palpable abdominal mass. Preoperative imaging rarely contributes to the final diagnosis and shows a non-specific mass with peripheral enhancement. A preoperative biopsy is often non-diagnostic because P-RMS requires special staining not routinely performed. On pathologic examination polygonal large rhabdomyoblasts and muscle specific markers for myogen, desmin, SMA and vimentin are used to confirm the diagnosis. The only possible treatment for adult RMS is surgical resection of the tumor followed by chemotherapy and radiotherapy. Data have shown that adult RMS are chemo-sensitive tumors with response rates of about 75% [11,12]. The combination of vincristine, dactinomycin, and cyclophosphamide has been established as the standard therapy in patients with non-metastatic RMS [15]. Other combination therapies have included ifosfamide and etoposide, although there have been no improvement in outcome when ifosfamide was substituted for cyclophosphamide, or when etoposide and ifosfamide were substituted for dactinomycin and cyclophosphamide. Multivariate analyses indicate that age, histologic subtype, primary site location, stage, local control with surgery and/or radiation are significant predictors of survival in adults [13–16]. Age and differences in distribution of histopathologic subtypes result in diminishing chemosensitivity and lower tolerance for intensive therapy [17]. Disease that responded to chemotherapy was associated with a better metastasis free interval [18]. A study by Ogilvie shows that when combined with surgery and radiation therapy, chemotherapy using doxorubicin, ifosfamide, and vincristine yielded 55% overall and 64% disease free survival at 2 years [11]. The study also showed that patients with localized disease who achieve a pathologic response by neoadjuvant chemotherapy can become disease free after surgery and radiation therapy. However, this study also showed that patients with metastatic disease fail to achieve a pathologic response due to other metastatic lesions not being

amendable to surgery. Long-term prognosis of P-RMS reported (predominantly of limbs) is poor and survival rates vary from 12.5 to 50 % in the 1-year following treatment in adults [19]. In our case the patient died 2 years following treatment due to recurrence of the disease. Further studies are needed in the adult population to establish an optimal therapy.

4. Conclusion

RMS is an extremely rare tumor in adults, but it should be included in the list of differential diagnosis of patients presenting with an atypical pancreatic or liver mass on imaging studies. Special staining for RMS should also be considered for the abdominal mass specimens to expedite the time to final diagnosis and starting the treatment.

Conflict of interest

Ali Shirafkan, and the other co-authors have no conflict of interest to declare.

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Consent

Written informed consent was obtained from the patient's next of kin for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Authors contribution

All authors have contributed in patient care, data collection, data analysis and interpretation and writing the paper.

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