is complete surgical resection. Tumor designated as high risk should be treated with adjuvant Imatinib (tyrosine kinase inhibitor). Imatinib is the mainstay of treatment for metastatic or unresectable tumor. Tumor resistant to Imatinib is treated with Sunitinib.

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Primary extraskeletal osteosarcoma of gall bladder

Introduction

Extraskeletal osteosarcoma (ESOS) is an uncommon malignant mesenchymal neoplasm constituting 1% of all soft tissue sarcomas[1] and 2-4 % of all osteosarcomas (OS). Though the primary skeletal OS is a common bone tumor of the first andsecond decades, ESOS has been reported in the elderly (60-70 years).[1] Allan's criteria for diagnosis of ESOSinclude: the tumor, (1) to be ofsoft tissue origin without any attachment to bone/periostium, (2) to have a uniform sarcomatous morphology without carcinomatous area and (3) shouldproduce malignant osteoid, bone or cartilaginous matrix.[2] The present article reports a primary ESOS of gall bladder (GB) in a 72 year old female . To the best of our knowledge this is the second case of the primary ESOS of the GB.[3] A written informed consent was taken in the index case.

Case report

A 72 year old diabeticfemale with coronary artery disease, bronchial asthma, hemiparesis and depression,presentedwith off and on pain on the right hypochondrium, radiating to the ipsilateral sub-scapular region for last 3 years,with loss of appetite and weight (lost10 kgsover 10 months). She had jaundice 2 years back, which resolved spontaneously.Onlocal examination and contrast enhanced computer tomogram (CECT) a distended GB with heterogeneous soft tissue within the lumen anddilated common bile duct (CBD) was noted without calcification or extra-luminal extension (**Figure 1A**). Magnetic resonance cholangiopancreatography (MRCP) also showed similar filling defects in the GB and lower CBD (**Figure 1B**). Radiologically, possibilities considered were: empyema with sludge and calculi or hydatid cyst with rupture. Carcinoma was not considered, as no definite wall thickening or soft tissue extension seen. Ultrasound guided fine needle aspiration cytology (FNAC) showed a few highly atypical cells, suggestive of a malignant tumor. The patient underwent cholecystectomy with partial CBD excisionas it was adherent to the duodenum and on per-operative frozen section examination a positive CBD margin was identified. No lymph node was identified. Grossly a polypoidal friable growth measuring 4.5x4x0.2 cms was identified near fundus of GB, involving the CBD, with attached liver bed measuring 3.2x1.5cms and a free liver margin of 2.1 cms. Microscopically, a pleomorphic malignant tumor infiltrating the wall of GB with overlying normal epithelial lining was seen. Within the tumor, numerous osteoclastic giant cells were noted with significant nuclear pleomorphism and foci of osteoid formation by the malignant cells, as confirmed by Verhoeff-Van Gieson stain (arrows) (**Figure 2A& 2B**). The proximal CBD margin was also involved by the tumor cells. Immunohistochemistry (IHC) showed positivity for

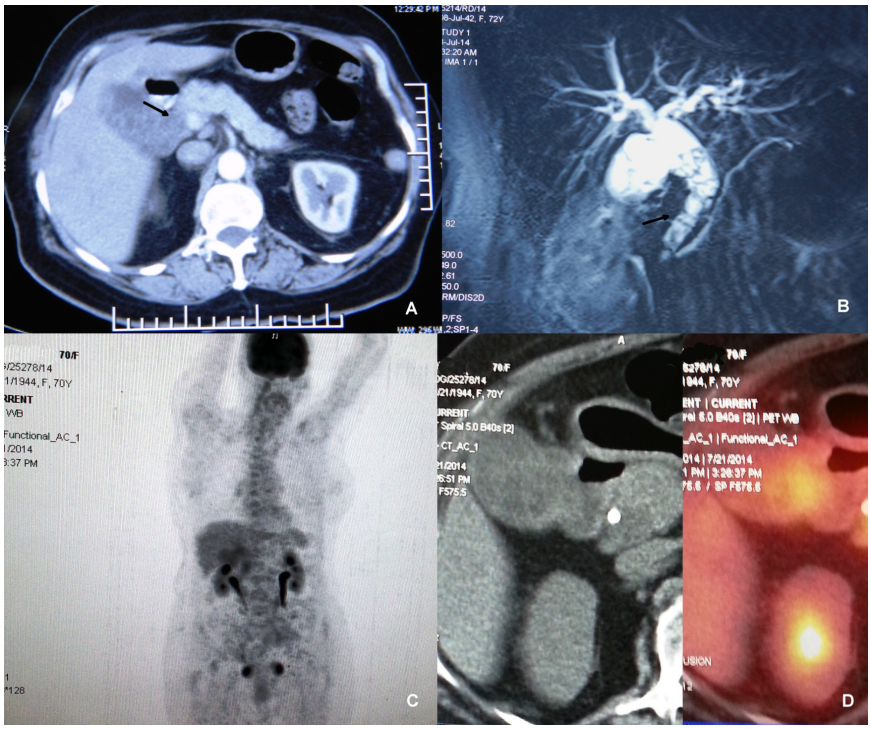

Figure 1: (A) Axial contrast enhanced CT scan image shows distended gall bladder with heterogeneous soft tissue within the lumen (arrow) without calcification. No extra-luminal extension is seen. (B) Magnetic Resonance CholangioPancreatography (MRCP) image shows a heterogeneous septate lesion in the gallbladder with similar appearance in the lower CBD causing dilatation of intrahepatic biliary radicles. No soft tissue component is seen. (C) Coronal PET image showing only mild uptake of FDG in the gall bladder. (D) CT and PET-CT fusion image showing the gallbladder lesion with mild FDG uptake.

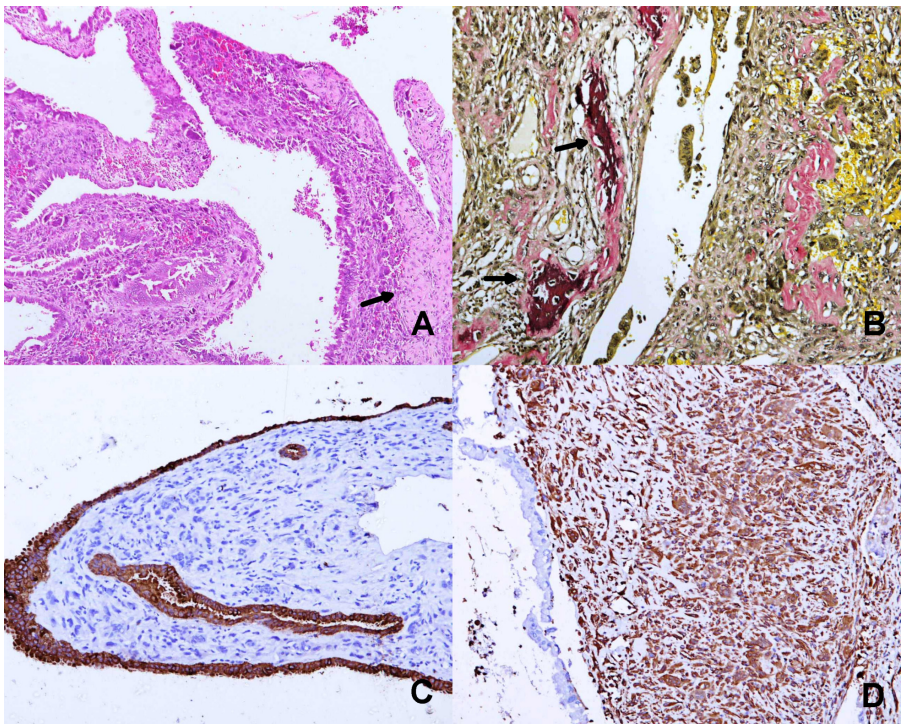

Figure 2: Photomicrograph shows aosteoclastic giant cell rich malignant tumor in the gallbladder wall with overlying non-dysplastic columnar epithelial lining. An island of osteoid formation (arrow) is seen [Figure 2A, H&E x100]. VVG stain confirms presence of tumor osteoid (arrows) [Figure 2B, H&E x100]. Cytokeratin stain show positive overlying columnar epithelium with submucosal positive glands. The tumor cells are negative [Figure 2C, IHC (CK) x100].Vimentin stain however shows diffuse cytoplasmic positivity in the tumor cells, while the epithelium is negative [Figure 2D, IHC (Vim) x100].

osteopontinand vimentinin the tumor cells andnegativity for pan-CK, CK19, myogenin and desmin (**Figure 2C & D**). The overlying epithelium was, however positive for CK. Histopathological (HPE) diagnosis of an ESOS (osteoblastic variant) of GB was made. Postoperative period was uneventful and positron emission topography (PET) showed no systemic uptake and she was further advised for close follow up (**Figure1C & D**).

Discussion

ESOS was first described by Wilson in 1941. Primary ESOS is a rare high grade sarcoma that arises from deeper soft tissueswith male predominance (1.9:1). The exact etiologyis unknown, however, two theories have been described: (a) tissue residue theory (embryogenic mesoblastic rest) and (b) metaplastic theory (of the interstitial fibroblastsby radiation exposure, trauma, surgery, etc.).[4]

As radiological features are nonspecific, a HPE examination is mandatory for diagnosis.[4,5] Six histological subtypes, similar to those of the skeletal OS are: (1) osteoblastic, (2) fibroblastic, (3) chondroblastic, (4) teleangectatic, (5) small cell and (6) well differentiated Os.In the index case the diagnosis of osteoblastic ESOS was made due to the presence of abundant osteoidina tumor with histology similar to the classical OS. Amongst the intra-abdominal ESOS, kidney, followed by the retroperitoneum and mesentery are other reported sites. Differential diagnosis are benign myositis ossificans, mature teratoma or calcified paragangliomas. Amongst the malignant tumors, undifferentiated giant cell rich carcinoma, sarcomatoid carcinoma with osteoclast like giant cells, carcinomsarcoma, dedifferentiated liposarcoma, extra skeletal mesenchymal chondrosarcoma, malignant mesenchymoma and malignant fibrous histiocytomas are to be considered in GB.[4,6,7] The "reverse zoning phenomenon"of ESOS, i.e. centrally placed osteoid and peripheral spindle cell proliferation, can differentiate from the "zoning phenomenon" of myositis ossificans (periphery with most mature component).[6] Histologically this was a malignant tumor and lack of CK positivity in the tumor cells, with vimentin expression and negativity for other markers made us to rule out carcinosarcoma or sarcomatoid carcinomas. De-differentiation was not identified. Considering all features, a diagnosis of ESOS was confirmed. To the best of our knowledge, this is the second report of this entity.[3]

The treatment ofchoice in ESOS is wide/ radical/ simple surgical resection, depending on the location and extent of lesion. However, high rates of recurrence (45%) and metastases (65%) results in short median survival time of 2 years, with 5-year overall survival rate of <37%.[4,9] Other known prognostic factors are: tumor size>5 cms in diameter, elderly age group and serum alkaline phosphatase levels. Lung (60%), followed by the regional lymph nodes, heart, bone and soft tissuesare the most preferred site for metastases of ESOS.[4,9] Though efficacy is not fully established, the survival rate may be improved by post-operative radiotherapy or chemotherapy, especially in young adults, as seen in well differentiated ESOS, with a reduced recurrence rate of 28%, in comparison to the 48% recurrence in surgically excised tumors without radiotherapy.[10] As rarely reported, there is no prognostic or therapeutic data available for the primary ESOS of GB. Though in the present case, the CBD margin was positive, however, in view of patient's old age and other co-morbidities, the patient was kept on close follow up and further not subjected to adjuvant treatment.

Conclusion

Primary ESOS of GB is an extremely rare tumor,with its predilection for elderly patients and atypical radiological findings. However, intra-abdominal ESOS is not so uncommon; so in an elderly patient with atypical radiological features and aggressive clinical course, this tumor should be considered in the differential diagnoses. Possibility of metastasis should always be ruled out and individualistic treatment should be tried, to build knowledge on these tumors.

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Endotipsitis caused by extremely drug-resistant *Klebsiella pneumoniae*

Introduction

Transjugular intrahepatic portosystemic shunt (TIPS) is commonly used for the decompression of portal hypertension. TIPS infection, also known as 'endotipsitis', is a rare but serious complication of TIPS insertion.[1] To date approximately 42 cases of 'endotipsitis' have been described. To the best of our knowledge this is the first case report of endotipsitis caused by an extremely drug-resistant Klebsiella pneumoniae (XDR).[2]

Case report

A 28-year-old man with decompensated chronic alcohol-related liver disease was admitted with upper gastrointestinal bleeding. He had no history of hospitalization in the previous 6 months. He underwent TIPS insertion due to variceal bleeding which did not respond to endoscopic variceal ligation. Three days post TIPS procedure he developed ARDS with leukocytosis, hyperbilirubinemia, elevated aminotransferases, hypoalbuminemia, coagulopathy and septic shock. Two sets of blood cultures grew extremely drug resistant (XDR) *Klebsiella pneumoniae* (resistant to all antibiotics tested including carbapenems, colistin, and tigecycline). Workup, including urine culture, transthoracic echocardiography, chest radiography, abdominal imaging, and diagnostic paracentesis, failed to localize the source of the bacteremia. He initially improved with a combination of multiple antibiotics (imipenem, colistin, tigecycline, chloramphenicol, amikacin, ciprofloxacin and fosfomycin). However he had re-bleeding: Doppler ultrasound did not show any thrombus and CT angiogram revealed left gastric artery bleeding for which embolisation was performed. His TIPS was re-examined via transjugular route to look for any thrombus. Stent patency was confirmed; however in view of the recurrent bleed, TIPS stent dilatation with balloon was done. After transient improvement he again developed fever, worsening of sepsis and persistent XDR

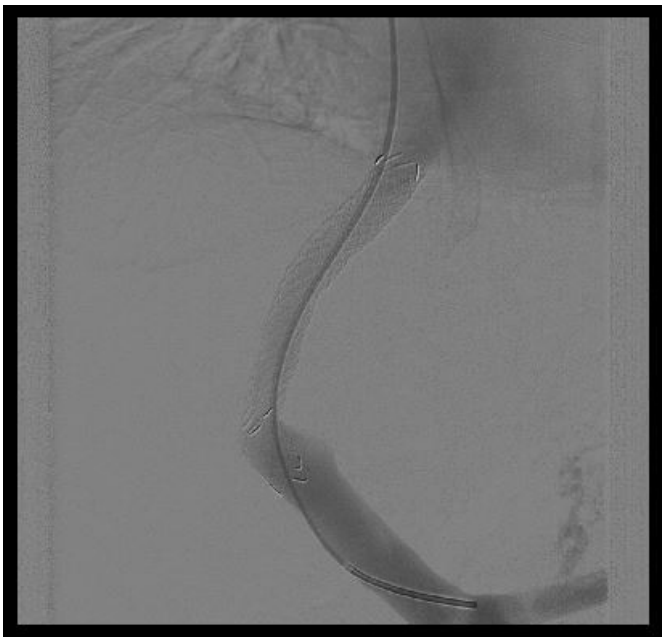

Figure 1: TIPS seen following contrast injection into the portal vein during the interventional proceedure (TIPSogram)