Primary duodenal extraskeletal osteosarcoma-a case report

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Abstract

Extraskeletal osteosarcoma is a rare malignant soft tissue tumour, which represents about 4% of all osteosarcomas and 1% of all soft tissue sarcomas. We present a case of extraskeletal osteosarcoma, albeit at a hitherto undescribed site, duodenum. This case report addresses the clinicopathological features and differential diagnosis of extraskeletal osteosarcoma and the importance of clinicopathological correlation in the diagnosis and workup of such cases.

Keywords: Extraskeletal osteosarcoma, duodenum, malignant soft tissue tumour, clinicopathological features

Introduction

Extraskeletal osteosarcoma is a malignant mesenchymal neoplasm that depicts malignant osteoid, bone and/or chondroid material. These tumours have no attachment to bone or periosteum, which has to be determined by the examination of radiological findings or observation during surgery [1,3]. It most commonly manifests in individuals older than 50 years with the lower extremity being the most common anatomic site, followed by the upper limb and the retroperitoneum [1,4]. We present a remarkable case of primary duodenal extraosseous osteosarcoma.

Case presentation

The patient was a 62 year old man who was admitted with history of intermittent pain abdomen of 3 years duration. There was no history of fever, vomiting, loss of appetite, loss of weight, diarrhoea or anti-tuberculosis treatment. Patient had no history of trauma or radiation therapy. Ultrasound scan abdomen showed a heterogeneous spherical mass measuring 13×11×9 cm arising from posterior wall of duodenum overlying renal hilum with central calcification occupying 20% of total area and eccentric calcification with irregular spiculated margins. Computed tomography of the abdomen revealed a large bulky mass involving duodenal and paraduodenal region with cholelithiasis (Figure 1). Endoscopic findings revealed a submucosal mass with superficial ulceration of duodenal mucosa. A biopsy performed was reported as a spindle cell lesion. Patient underwent Whipples' pancreaticoduodenectomy.

Intraoperative findings showed a large mass arising from 2nd and 3rd part of duodenum and on palpation appeared solid and cystic. Mass was adherent to inferior vena cava, Gerotas' fascia and perinephric fat on the right side. Macroscopic examination revealed a sub-mucosal duodenal mass measuring 11×9×8 cm. Cut surface appeared solid-cystic, with cysts of varying sizes containing haemorrhagic fluid. Focal firm to hard calcified areas were seen within the lesion (Figure 2).

Microscopic examination revealed a cellular tumor involving the sub mucosa, muscularis propria and subserosa of the small intestinal wall with superficial mucosal ulceration. The tumor cells were arranged in short interlacing fascicles of spindle to epithelioid cells. The cells exhibited mild to moderate pleomorphism having eosinophilic cytoplasm, vesicular nuclei with prominent nucleoli. Mitosis of 32/50 HPF were noted. Areas of haemorrhage and dilated vascular channels were seen. Areas of osteoid were seen between the tumor cells with scattered osteoclastic giant cells (Figures 3A and 3B). Foci of dystrophic calcification were noted. Adjacent pancreatic tissue was infiltrated by the tumor. Immunohistochemistry was performed; the tumor cells were positive for vimentin, osteonectin, CD 68 and negative for CK, desmin, cKit, S100, CD34 and SMA (Figures 3C and 3D). A diagnosis of extraskeletal osteosarcoma was made. The patient had an uneventful post operative period and remains free of tumor a year after surgery.

Discussion

Extraskeletal osteosarcoma is an uncommon tumor and there

Figure 1. Heterogeneous mass with calcification.

Figure 2. Submucosal duodenal mass with solid-cystic cut surface.

have been less than 50 cases reported so far [5]. Usually affects adults with a high incidence in patients older than 50 years and is slightly more common in males than in female patients [1]. The lower extremity is the most common site followed by upper limb and retroperitoneum [4,6]. Unusual sites, such as the larynx, kidney, oesophagus, small intestine, liver, heart, urinary bladder, parotid, and breast have been described [4,6]. The pathogenesis of the tumor is unclear, but prior history of radiation [4], a previous history of trauma at the tumor site and malignant transformation of myositis ossificans to extraskeletal osteosarcoma have been proposed [4].

Figure 3. Tumor cells during mitosis and after immunohistochemistry analysis.

- (A) Cellular lesion composed of tumor cells [H&E, ×4].
(B) Tumour cells with osteoid and osteoclastic giant cells [H&E, ×40].
(C) Immunoreactivity of tumour cells to anti-osteonectin [H&E, ×40].
(D) Immunoreactivity of tumour cells to anti-vimentin [H&E, ×40].

The histological patterns are varied as similar to osteosarcomas, which includes osteoblastic, fibroblastic/pleomorphic malignant fibrous histiocyte like, chondroblastic, giant cell MFH like, small cell, mixed and telangiectatic pattern. Nevertheless, the major predictor of clinical outcome was found to be the tumour size (>5cm) according to the study by Bane et al [4]. According to ED B Chung and F M Enzinger, extraskeletal osteosarcoma with a prominent fibroblastic or MFH like component seemed to have a slightly better prognosis than predominantly osteoblastic, chondroblastic or telangiectatic types of the tumour [5]. The biological behaviour of well differentiated extraskeletal osteosarcoma has been suggested to be better than that of classical extraskeletal osteosarcoma, but some cases can progress to a higher grade [7]. It is necessary to distinguish extraskeletal osteosarcoma from other benign or malignant bone and cartilage forming soft tissue lesions. Amongst the malignant tumours, metaplastic bone is often seen in synovial sarcoma, malignant fibrous histiocyte, dedifferentiated liposarcomas with osseous or cartilaginous elements, malignant peripheral nerve sheath tumour with heterologous elements, malignant melanoma and carcinosarcoma. Adequate sampling may be necessary to keep us from making an erroneous diagnosis. Extraskeletal osteosarcoma has to be distinguished from myositis ossificans which shows a characteristic zoning phenomenon with peripheral differentiation into well formed bone and a lack of cytologic atypia. A "reverse zoning phenomenon" with central osteoid material deposition and atypical cell proliferation at periphery is characteristic of sarcoma [4].

Elevated alkaline phosphatase levels has been accepted as a prognostic tumour marker in osteosarcomas [8], albeit, such high levels of alkaline phosphatase have also been reported in extraskeletal retroperitoneal osteosarcomas and is unlikely to be found in other retroperitoneal soft tissue tumours [9].

The immunohistochemical profile of vimentin, osteonectin, CD 68 positivity and CK, desmin, cKit, S100, CD34 and SMA negativity supported the cytomorphology and favoured a diagnosis of extraskeletal osteosarcoma, as in our case. Osteocalcin and osteonectin help to distinguish malignant bone from collagen and cartilage matrix which is essential to the diagnosis of extraskeletal osteosarcoma [10]. To distinguish between low grade osteosarcoma and benign lesions, molecular analysis may be helpful that shows amplification of SAS, CDK4 and MDM2 genes in 12q 13-15 region in the former lesions [11].

According to a study by Lee JS and et al., patients with extraskeletal osteosarcoma generally have a poor prognosis and the majority develops metastatic disease within 3 years of diagnosis [12,13]. It has been reported that the overall mortality rates due to the tumor in the larger series exceeded 60% [1,2,5]. Radical resection appears to be the best therapeutic option for local control but has no effect on distant metastasis [14]. Due to the rarity of cases, the efficacy of chemotherapy has not been evaluated although it has been the commonly used treatment modality [15]. Temporary palliative therapy includes radiotherapy [16], however understanding of the tumour biology and gene over-expression is required to enable efficient treatment.

Conclusion

Our case is illustrative of the well described extraskeletal osteosarcoma at an unusual site, duodenum. There is a need for a thorough sampling of the surgical specimen for documenting the presence of malignant osteoid and to exclude other differential diagnosis. Higher degree of accuracy in the diagnosis of such lesions requires clinical, radiological, histological and immunohistochemistry correlation.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

Authors' contributions	HN	AK
Research concept and design	✓	✓
Collection and/or assembly of data	✓	--
Data analysis and interpretation	--	✓
Writing the article	✓	--
Critical revision of the article	✓	--
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Statistical analysis	--	--

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