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Extraskkeletal Ewing's Sarcoma Of The Small Bowel

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Abstract: *Extraosseous Ewing's sarcoma also referred to as Extraskkeletal Ewing's sarcoma is a rare tumor of primitive cells which occurs in children, adolescents and young adults. This tumor mainly involves the soft tissue of the extremities and thorax. We report a case of Extraskkeletal Ewing's sarcoma of jejunum without associated skeletal location. A 22 year old female presented with complaints of pain abdomen and vomiting for two days. CT report was leiomyosarcoma /GIST arising from one of the distal jejunal loops. After surgery the specimen was sent to the pathology department. We received 35cm long intestinal segment showing 10x8x6cm grey brown nodular mass, which on cut section extending into the luminal aspect of small intestine. On histopathological examination it was reported as small round cell tumor. The tumor is immunoreactive for CD99 & S100 protein and immunonegative for CD45. Based on these features it was reported as Extraskkeletal Ewing's sarcoma.*

Keywords: *Extraskkeletal Ewing's sarcoma, Small Bowel, Jejunum.*

I. INTRODUCTION

Ewing's sarcoma is a rare tumor of primitive cells which is located mainly in the spine and chest wall, in lower extremities and retroperitoneum.^[1,2] These cases also have been documented in the pancreas,^[3,4] vagina,^[5] rectovaginal sectum,^[6] prostate,^[7] ovaries,^[8] oesophagus,^[9] kidney^[10] and stomach.^[11] Cases of small bowel Extraskkeletal Ewing's sarcoma were also reported.^[12,13] We describe this case because of its rarity.

II. CASE REPORT

22 years old female came with pain abdomen and vomiting for two days. Ultra sound examination revealed hypoechoic mass suggestive of retro peritoneal mass. CT examination showed a large heterogeneously enhancing exophytic mass

involving left lumbar region, arising from one of the distal jejunal loops and reported as leiomyosarcoma /GIST. After surgery the specimen histopathology laboratory .

A. Gross Appearance

The received specimen was 35cm long intestinal segment with 10x8x6cm grey brown nodular mass along the anti mesentic border (fig-1). Cut-section revealed solid grey white and dark brown mass extending into the luminal aspect of the intestine (fig-2).



Fig.1:



Fig.2

B. Microscopic appearance

H&E stained sections show sheets of uniform small round cells with scant cytoplasm and round to oval vesicular nuclei with stippled chromatin in some of them. These cells are extending from mucosa to serosa and are separated by areas of necrosis with perivascular localization and pseudorosette formation at places(fig-3,4,5). Mesentery shows few congested blood vessels and small clusters of tumor cells. Based on these features it was reported as small round cell tumor. After that immunohistochemical examination was done and the tumor is immunoreactive for CD99(fig-7). & S100(fig-8). It is immuno negative for CD45(fig-6). Based on these features it was diagnosed as Extraskelatal Ewing's sarcoma.

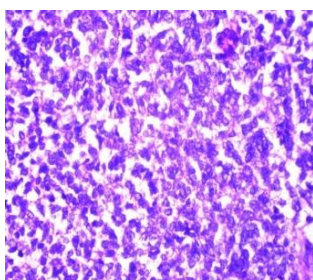


Fig-3

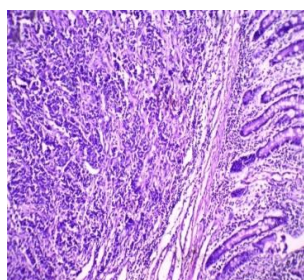


Fig-4

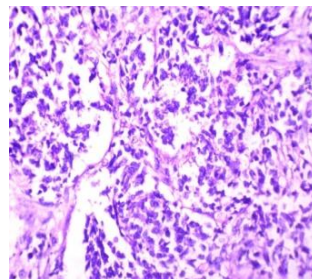


Fig-5

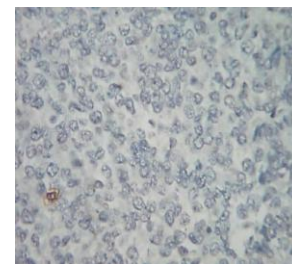


Fig-6

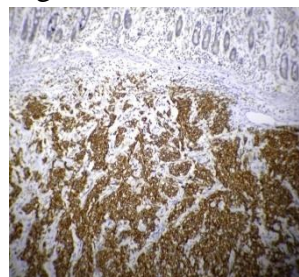


Fig-7

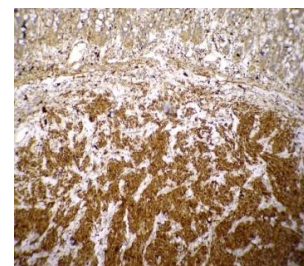


Fig-8

III. DISCUSSION:

Ewing's sarcoma(ES) was first discovered by James Ewing in 1921 as diffuse endothelioma of bone. The Ewing's family of sarcomas includes Ewing's sarcoma of bone, Extrasosseous Ewing's sarcoma is also referred to as Extraskelatal Ewing's sarcoma, primitive neuroectodermal tumor, peripheral neuroepithelioma, askin's tumor(Ewing's sarcoma of chest wall) and atypical Ewing's sarcoma. In 90% of cases ES family of tumors is found in patients between 5&25 years of age. After age 25 it is exceptionally rare, demonstrating aggressive behavior and frequent recurrence.^[14] Dickinson et al. found a prevalence of 0.2 cases/million inhabitants.^[15] In Ewing's sarcoma the translocation is between chromosomes 11&22 and is refered to as t(11;22). The new fused gene, called EWS/FLI.^[14] Histologically these tumors are composed of small round cells that are rich in glycogen and the neuroepithilial differentiation in the form of pseudorosette. The neuro endocrine phenotype is confirmed by immune positivity to CD99 and to NSE &S100 to a lesser extent as these are also found in number of other small cell tumors.^[8,16] Treatment is surgery combined with chemotherapy and high dose radio therapy for favourable outcome and low incidence of recurrence.^[17]

IV. CONCLUSION

Extraskelatal Ewing's sarcomas are a group of malignant tumors having poor prognosis. The diagnosis of Extraskelatal Ewing's sarcoma is challenging when evaluating small bowel tumors. Examination of H&E stained sections is a gold standard, however Immunohistochemistry and cytogenetic analysis are essential for diagnosis of Extraskelatal Ewing's sarcoma.

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