Case Report

Pathology Section

The Cytology of the Benign Extra-Gastrointestinal Stromal Tumour in the Pouch of Douglas: A Case Report

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ABSTRACT

Extra-gastrointestinal stromal tumours are rare, non-epithelial, mesenchymal tumours which arise from the soft tissues of the abdomen- mesentry, the omentum and the retroperitoneum. These tumours are histologically and cytologically similar to the stromal tumours of the gastrointestinal tract, they are composed of purely rounded epithelioid cells or short fusiform cells or a mixture of both and are set in a fine fibrillary myxiod background. The cytological features of the imprint smears in a case of benign extra-gastrointestinal stromal tumour of the mesentry, projecting into the pouch of douglas in a 42-year old female, have been described here. The recognition of these tumours is important because of their aggressive biological behaviour. The metastatic potential and the high rate of recurrence of these tumours necessitate the frequent follow up of the patients after a surgical resection.

Key Words: Epithelioid, extra-gastrointestinal, spindle, stromal tumour

INTRODUCTION

Extra-gastrointestinal stromal tumours (EGISTs) are very rare tumours, accounting for less than 10% of the stromal tumours which arise in the gastro-intestinal tract [1]. They constitute a group of primary non-epithelial mesenchymal tumours which arise outside the gastrointestinal tract in the soft tissues of the abdomen like the mesentry omentum and the retroperitoneum, with the exclusion of the tumours which arise in the gastro-intestinal tract and the exclusion of those having the classic features of leiomyoma, leiomyosarcoma, schwannoma and other fibromatoses. These tumours are believed to arise from the primitive stromal cells which are capable of differentiating into intestinal smooth muscle or neural cells, because some of these tumours show positivity for the smooth muscle and neural markers.

The cytology of a case of benign extra-gastrointestinal tumour in a 42-year-old female has been presented here.

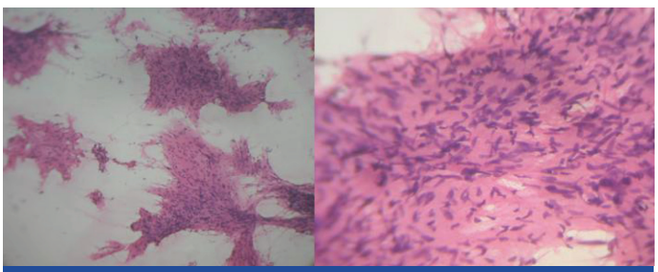
CASE REPORT

A 42-year-old, premenopausal, multiparous female presented with lower abdominal pain of 3 months duration. The general physical examination was normal. The per-abdominal examination revealed no palpable mass, except for tenderness in the right iliac fossa. The per-vaginal examination showed a tender right fornix, a bulky uterus and fullness in the pouch of douglas. The other systems were normal. Ultrasound examination of the lower abdomen showed a right sided pyosalphinx and a hypoechoic mass in the posterior aspect of the uterus. Abdominal hysterectomy with bilateral salphingo-oopherectomy was done. Per-operatively, a solid tumour which measured 6×4 cms, which arose from the mesentry and was protruding into the pouch of douglas, was made out. This mass was not attached to the gastrointestinal tract or to the genitourinary organs. The mass was enucleated and sent separately for histopathological examination.

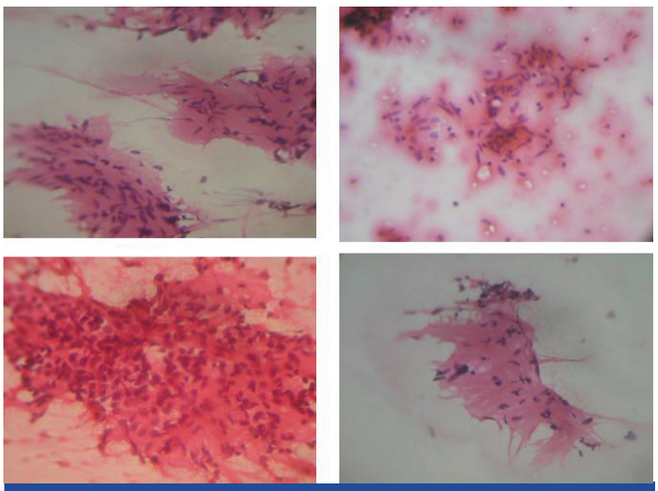
The mass was well circumscribed and unencapsulated. The cut section showed a smooth, lobulated and tanned appearance with solid, cystic and haemorrhagic areas. Imprint smears of the unfixed specimen were taken, fixed in ethyl alcohol and stained with the hematonylin- eosin (H and E) stains.

The cytology revealed many tissue fragments which consisted of loose spindle cells in a myxoid fibrillary background. These spindle cells which had ill-defined cytoplasm were arranged as fascicles or palisades, with spindle or cigar shaped nuclei, with pointed or blunt ends, with finely dispersed chromatin and indistinct nucleoli [Table/Fig-1 and 2a]. Areas showing loosely cohesive sheets of oval or polygonal cells with moderate, well defined cytoplasm and round hyperchromatic nuclei with granular chromatin and nucleoli, which represented an epithelioid morphology were seen [Table/Fig-2c]. Extra cellular, amorphous eosinophilic material with few spindle cells, which represented skenoid fibres [Table/Fig-2d] and many stripped nuclei [Table/Fig-2b] were seen. The diagnosis of stromal tumour of extra-gastrointestinal origin was suggested.

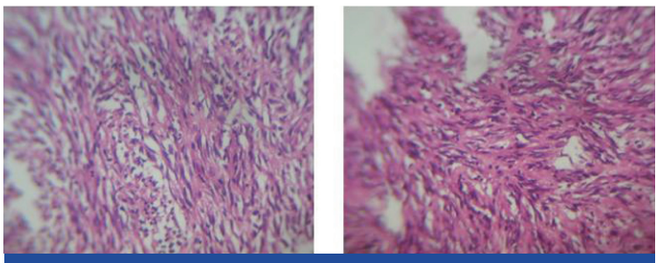
The haematoxylin and eosin stained paraffin sections showed interlacing fascicles of benign spindle cells with eosinophilic fibrillary cytoplasm and cigar shaped nuclei. There was no nuclear pleomorphism, mitoses or necrosis. A histopathological diagnosis of extra-gastrointestinal stromal tumour was made and it correlated with the cytology. Immunohistochemistry of the tumour showed positivity for CD [1].



[Table/Fig-1]: Smears showing tissue fragments consisting of spindle cells in a myxoid fibrillary background. [H and E, X100 (Left), ×400 (Right)]



[Table/Fig-2]: Smears showing
(a): spindle cells with spindle nuclei with pointed ends (upper left, H and E, × 400)
(b): stripped fusiform nuclei (upper right, H and E, × 400)
(c): epithelioid cell morphology (lower left, H and E, × 400)
(4): skenoid fibres (lower right, H and E, × 400)



[Table/Fig-3]: Sections showing interlacing fascicles of spindle cells with eosinophilic fibrilllary cytoplasm and wavy nucleus (H and E ,× 400)

DISCUSSION

Stromal tumours which arise outside the gastro-intestinal tract are very rare [1]. These tumours arise from the soft tissues of abdomen-mesentry, the omentum and theretroperitoneum. EGISTs have been extensively analyzed by Reith et al [1]. EGISTs have been documented in various sites like the omentum, the pancreas and the vulvoviginal and the rectovaginal septa [2] .

Extra-gastro-intestinal stromal tumours (EGIST) are cytologically and histologically similar to gastro-intestinal stromal tumours which are cellular spindle cell or epithelioid tumours which express the CD 34 and the CD 117 (c-kit) antigens [5]. EGISTs display various lines of differentiation which reflect the elements of the gut wall, showing differentiation towards smooth muscle and neural elements, dual differentiation and those that lack differentiation towards either cell type [6].EGISTs occur commonly in adults, presenting with abdominal pain or are discovered incidentally during a work up for an unrelated condition [1].

Grossly, these tumours tend to be lobulated, nodular, well circumscribed and unencapsulated with a smooth fleshy, whorled-silk appearance on cut sections. Areas of necrosis and haemorrhage are common in the malignant tumours [6].

Morphologically, EGISTs are sub-classified to be of the spindle or epithelioid types [5]. The cytology of these tumours have been described by Mills and Contos [7]. The aspirate smears are moderately cellular and are composed of both tight three- dimensional aggregates and noncohesive single cells which are dispersed in a relatively clean background. The nuclei are spindle shaped or epithelioid with a smooth nuclear membrane and they are evenly distributed, with finely granular chromatin and inconspicuous nucleoli. The cytoplasm is cyanophilic, delicate and fibrillar, with long tapered ends. Extra-cellular fibrillary matrix material is usually present. Skenoid fibres, which are not common markers in EGISTs, were found in our case. Additional morphological variations like a prominent myxoid matrix, a signet ring cell, granular cell features and oncocytic cytoplasmic features have been described [6].

The spindle type of EGISTs should be differentiated from solitary fibrous tumours, fibromatosis, inflammatory fibroid tumours, schwannomas, leiomyomas and leiomyosarcomas [6]. The epithelioid type of EGISTs should be distinguished from carcinomas, neuroendocrine tumours, melanomas and hepatocellular carcinomas [8].

The cytological features which predict the adverse outcomes are mitotic activity, cellularity and necrosis. Cytology can be used to diagnose EGISTs, but it is not reliable for assessing the malignant potential [9]. A histopathological examination is mandatory for all cases of EGISTs.

EGISTs are an aggressive group of stromal tumours with a malignant potential and a high rate of recurrence [1, 4]. But our case had a benign cytology. The malignant EGISTs metastasize to the lung, liver and other organs [6].

EGISTs are treated by surgical resection and the administration of tyrosine kinase inhibitors, with frequent follow ups to detect the recurrence [6].

To conclude, EGISTs are very rare aggressive tumours with high metastatic potential and a high recurrence rate. Cytology can be used as a confident diagnostic tool to detect these tumours. Histological examination is mandatory for the assessment of the malignancy to detect parameters like cellularity, mitoses, anaplasia, pleomophism and necrosis.

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