

Leiomyomatous Hamartoma of The Pyloric Antrum: A Case Report

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ABSTRACT

Leiomyomatous hamartomas are extremely rare lesions with very few cases being reported in the lungs and the oral cavity. There has been no published report of gastric leiomyomatous hamartoma till date. We present a case of leiomyomatous hamartoma in the pyloric antrum clinically mimicking a gastrointestinal stromal tumour. A 49-year old female presented with abdominal pain since 3 months. Endoscopy and Contrast-enhanced CT showed a large, well-defined, submucosal lesion in the antropyloric region, causing mild luminal compromise. In our case, distal gastrectomy (Bilroth 1) was done. Histopathological examination revealed a lesion comprising of proliferating connective tissue with smooth muscle fascicles dispersed in loose fibrous stroma, nerves and multiple small vessels. Immunophenotyping with smooth-muscle actin and desmin supported the diagnosis. We report this rare case and suggest the inclusion of leiomyomatous hamartoma in the differential diagnosis of gastric lesions with smooth muscle as a dominant component.

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Introduction

Magnus-Alsleben first described gastric hamartoma in 1903. [1] In 1904, Alberecht^[2] defined hamartomas as nonneoplastic malformations presenting as overgrowths of abnormal mature tissues, haphazardly arranged and indigenous to a specific site. The diagnosis is based on the histological appearance with smooth muscle bundles comprising as a dominant component along with multiple small vessels, and is confirmed by immunohistochemistry (IHC) with positivity for smooth muscle actin (SMA) and desmin.

Case Report

A 49-year old female presented with pain abdomen since 3 months. The pain was of moderate intensity, not related to food intake and was relieved by oral analgesics. There was no associated history of loss of weight or appetite. On clinical examination, the abdomen was soft and non-tender and there was no organomegaly. Endoscopy revealed a large submucosal lesion with central umbilication in the gastric antrum. Contrast enhanced CT scans confirmed a well-defined rounded mass lesion in the antropylic region, causing mild luminal compromise. A clinical diagnosis of gastrointestinal stromal tumour was made. Distal Gastrectomy (Billroth 1) was done.

The surgical specimens were formalin fixed and paraffin embedded. The sections were stained with routine hematoxylin and eosin stain. IHC staining was performed by the streptavidin-biotin-peroxidase method and diaminobenzidine as chromogen. The antibodies used included SMA, Desmin, CD117, S-100 and CD34. Appropriate positive and negative controls were performed.

Grossly, gastrectomy specimen was received measuring 9 x 5 x 3.5cm. A single globular lesion was identified in the submucosa measuring 5 x 3.5 x 2cm. A separate small nodule is identified in the attached omental tissue measuring 1 cm in diameter.

Microscopically, the lesion in the stomach comprised of loosely arranged smooth muscle fascicles with benign spindle-shaped cells having abundant eosinophilic cytoplasm and plump nuclei displaying minimal atypia. These fascicles were dispersed in loose fibrous stroma, nerves and multiple small vessels. Misplaced gastric glands were seen in the hamartomatous area which confirmed the diagnosis of a leiomyomatous hamartoma. However, no atypical changes were observed either in the epithelial or mesenchymal components. Additional investigations with Masson's trichrome allowed the red-stained smooth muscle cells to be clearly distinguished from the blue stained surrounding collagen fibres. Immunohistochemistry revealed a strong positivity of

smooth muscle bundles for SMA and Desmin and an immunonegativity for CD117. The lesion in the omentum showed only benign adipose tissue.

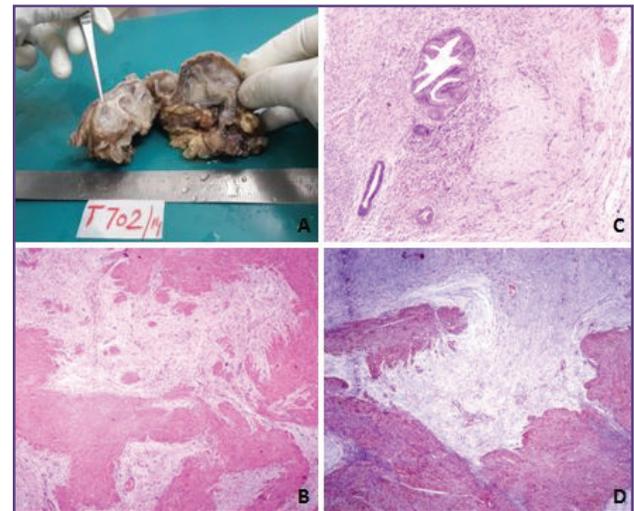


Fig. 1: (A) Gross photograph showing a submucosal lesion in the pyloric antrum. (B) Proliferating smooth muscle cells in a collagenous stroma intermixed with blood vessels (H&E, 4x). (C) Misplaced gastric glands in a hamartomatous area. (D) Smooth muscle fascicles scattered in a fibrous stroma (Masson's trichrome stain).

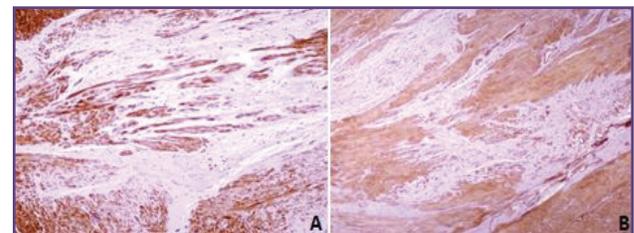


Fig. 2: (A) Smooth muscle bundles showing a strong positivity for smooth muscle actin (IHC) (B) Smooth muscle bundles showing a strong positivity for Desmin (IHC).

Discussion

Hamartomatous growth of the pyloric antrum is not common. They are rare benign tumours of the gastrointestinal tract. An abnormally arranged benign growth of cells normally found in that organ or site is defined as a hamartoma. Leiomyomatous hamartoma represents a 'true' hamartoma or abnormal growth with smooth muscle tissue as dominant component.

A few cases have been reported in the lungs and oral cavity. A review of the published reports regarding pulmonary and oral leiomyomatous hamartomas suggests that young females are predominantly affected with the most common age group being 0-8 years.³

The exact pathogenesis of this tumor is controversial. Some authors claim it to be metastases from a benign metastasizing leiomyoma or leiomyosarcoma, most often from the uterus,⁴ in which the tumor cells show monoclonal neoplastic proliferation of smooth-muscle cells; whereas others consider it as a primary smooth-muscle hyperplasia, tumor or hamartoma,⁵ showing polyclonal non-neoplastic proliferation of smooth muscle cells. Underlying dysgenic events that might affect the blood vessel formation may also lead to the development of leiomyomatous hamartoma.

Histologically, the lesion is comprised of a well-circumscribed nodule composed of smooth-muscle cells with numerous small vessels. In our case, a few misplaced gastric glands were seen entrapped within the smooth muscle component, with no atypical changes. The two conditions, benign metastasizing leiomyoma and a 'true hamartoma', or benign muscular proliferation, are indistinguishable from each other.⁶

Differential diagnoses include primary leiomyoma, primary leiomyosarcoma, metastatic leiomyosarcoma, fibroleiomyomatous hamartoma or benign metastasizing leiomyoma, lymphangiroleiomyomatosis and leiomyomatous hyperplasia.

Primary leiomyoma displays a characteristic whorling pattern and immunopositivity for SMA and Vimentin. Leiomyosarcoma shows frequent mitosis, nuclear atypia and is also immunoreactive for SMA and Vimentin. Fibroleiomyomatous hamartoma is seen mainly in lungs. There are whorled interlacing and communicating bands of smooth muscle which are separated by collagen, hyalinized stroma and fibrous tissue. The entity of lymphangiroleiomyomatosis (LAM) is seen rarely in lungs and kidney. In LAM lung disease there is abnormal proliferation of smooth muscle cells that line the lung airways and blood vessels. Leiomyomatous hyperplasia is characterized by abnormal proliferation of smooth muscles resembling benign leiomyoma but whorling pattern is not seen.

Previous published reports of oral and pulmonary leiomyomatous hamartoma have used SMA and desmin for confirmation of diagnosis. In the present case also, the tumor cells showed intense positivity for actin and

desmin. Additional markers like Collagen IV and CD34+ have been studied by few authors,⁷ showing collagen IV immunopositivity in the basement membrane of smooth-muscle cells.

We report this rare case and suggest the inclusion of leiomyomatous hamartoma in the differential diagnosis of gastric lesions with smooth muscle as the predominant component.

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Competing Interests

None Declared

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