

Diffuse Leiomyomatosis of the Esophagus

ÖZOPAGUSUN DİFFÜZ LEOMYOMA TOZİSİ

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SUMMARY

A case of diffuse leiomyomatosis of the esophagus in a 54 year old female patient is presented. The lesion gave a very long period of discomfort to the patient (19 years) and was cured with surgical resection of the esophagus along with proximal gastric segment. No associated pathology or familial clustering was observed in our case.

Key Words: Esophagus, Leiomyomatosis

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OZET

64 yaşında bir kadında belirlenen diffüz özofagus leomyomatozis olgusu sunulmuştur. Hasta 19 yıldır şikayet sahibiydi. Cerrahi tedavi edilen hastada yandaş patoloji ve familial eğilim belirlenmedi.

Anahtar Kelimeler: Özofagus, Leomyomatozis

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Leiomyomas are the most common benign tumors of the esophagus, and multiple lesions are rare. In a series of 838 reported cases of leiomyomas only 2.4% were multiple (1). Diffuse esophageal leiomyomatosis is much rarer, sometimes accompanying associated lesions or showing familial clustering (2-6). We report a case of diffuse leiomyomatosis which caused dysphagia for 19 years.

CASE REPORT

A 54 year old housewife was admitted to our hospital with a complaint of dysphagia for the last 19 years. She vomited food time to time. No family history of similar complaints were recorded. She had been formerly evaluated at several hospitals but she refused operation. Due to weight loss and increase in her complaints she was accepting a surgical intervention. Esophagogram revealed a large and dilated, twisted esophagus, poor emptying besides a 6 cm stenotic segment over cardia. Esophagoscopy showed a large

dilated lumen. A biopsy taken during endoscopy was evaluated as smooth muscle proliferation. Operation was undertaken through the transhiatal route: Resection of the thoracic part along with the abdominal portion of the esophagus and proximal stomach was performed. Reconstruction was accomplished by using stomach through esophageal bed. The patient had an uneventful postoperative course and was well one year after surgery without any complaint.

Pathology: The resected specimen measured 27 cm in length, weighted 1100 gm (Fig. 1). Distal portion was thickened up to 5 cm whereas mucosal nodularity was prominent in the proximal part. The cut surface of the lesions showed pallor a whorled appearance having pallor. Histologically the lesion consisted of nodules of smooth muscle fibers arranged in irregularly interlacing bundles without any mitotic activity (Fig. 2 and 3). The nodules were seen both in the proper muscle layer and muscularis mucosa.

DISCUSSION

Leiomyoma of the esophagus is mostly solitary and diffuse leiomyomatosis is extremely rare (2). Fernandes et al describes a case and reviewed 14 patients ranging 17 to 74 years of age with similar lesions. This lesion occurs approximately twice in males than females (3). Familial cases in association with AI-

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Fig. 1. Resected specimen showing the mucosal nodular appearance on the proximal segment and greatly thickened distal portion

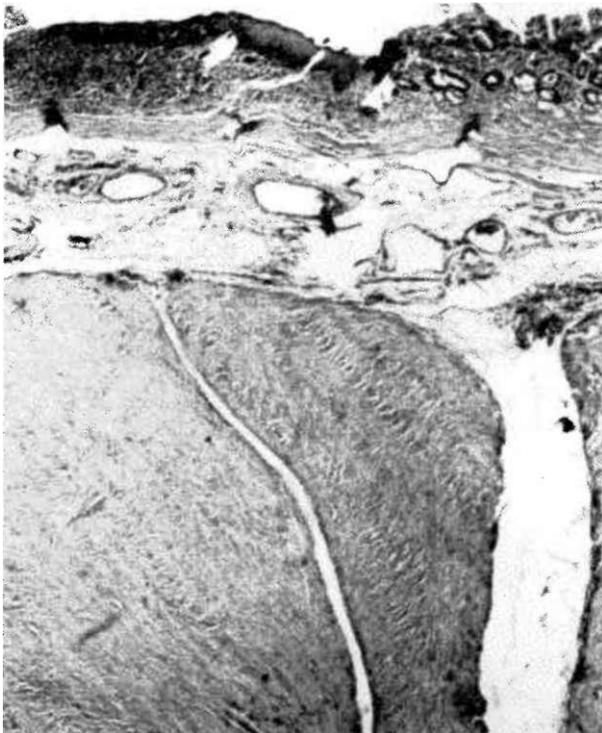


Fig. 2. Section through cardio-esophageal junction showing nodular growth beneath the submucosal layer (Hematoxylin and eosin, x, $\times 4$)

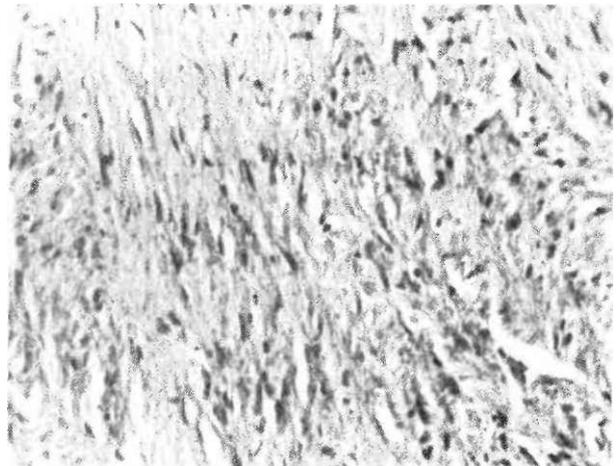


Fig. 3. High power view of the lesion showing spindle cells with smooth muscle features arranged in poorly defined bundles (Hematoxylin and eosin, x310)

port's syndrome (5) and also familial clustering with an autosomal dominant mode of inheritance associated with intestinal leiomyomas and neurofibromas along with urticaria pigmentosa or systemic mast cell disorders has been reported (4). Our case did not show any familial clustering and associated lesions.

Dysphagia can be present for a considerable period of time as was in our case. On the other hand cases may be without any symptom and found incidentally (3).

Our case is in accord with previous reports that accept the diagnosis of diffuse leiomyomatosis when the involvement is widespread along the esophagus with some nodularity rather than diffuse thickening of the muscular wall (1,2).

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