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*Clinical Image*

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# *Lumpy Bumpy Esophagus: A rare Cause of Pseudo achalasia*

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## Clinical image description

A 13-year-old girl presented with progressive dysphagia for 6 months. Computed tomography on Axial (Figure 1A) and Coronal plane (Figure 1B) showed a soft tissue mass in lower esophagus (white arrows), GE junction (red arrow) with proximal oesophageal thickening (arrow head). Biopsy of lesion showed leiomyoma. Due to progressive dysphagia esophagectomy with partial gastrectomy was performed. Specimen showed well-circumscribed firm nodules with overlying normal mucosa throughout the wall of distal esophagus and stomach. Microscopy revealed normal esophageal and gastric mucosa with underlying tumour arising from the inner layer of muscular is propria arranged in interlacing fascicles and whorls (Figure 2A). Tumour cells were spindly with blunt ends and bland nuclei without atypia or mitosis (Figure 2B). SMA (Figure 2C) and Desmin were strong diffuse positive on immunohistochemistry, suggestive of Diffuse Esophageal leiomyomatosis (DEL). CD117, CD34 were negative, ruling out gastrointestinal stromal tumour. MIB1 labelling index was <1% (Figure 2D). DEL is a smooth muscle benign hamartomatous lesion. A review study by Ziogas et al. in 2018 reported 35 cases under age of 18. It is commonly associated with Alport syndrome, but our patient had no symptoms of Alport syndrome. Histopathology including immunohistochemistry is the gold standard for diagnosis of DEL. Surgical resection is the treatment of choice in patients with severe, progressive symptoms.

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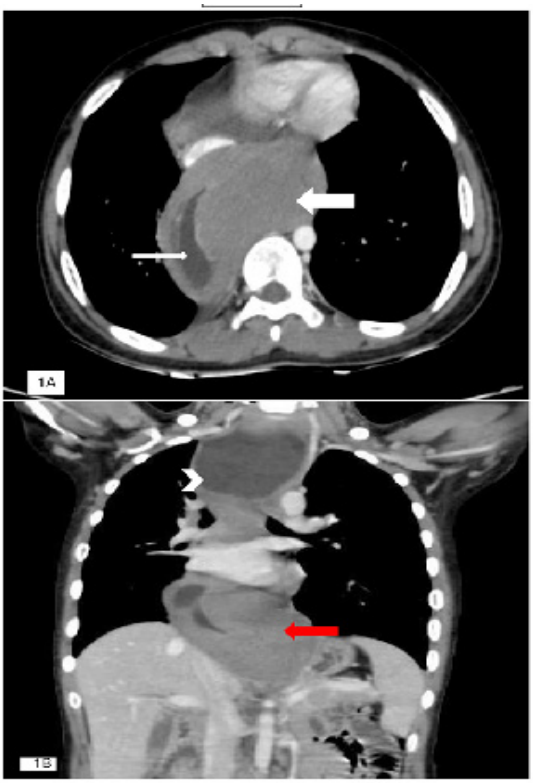


**Figure 1**

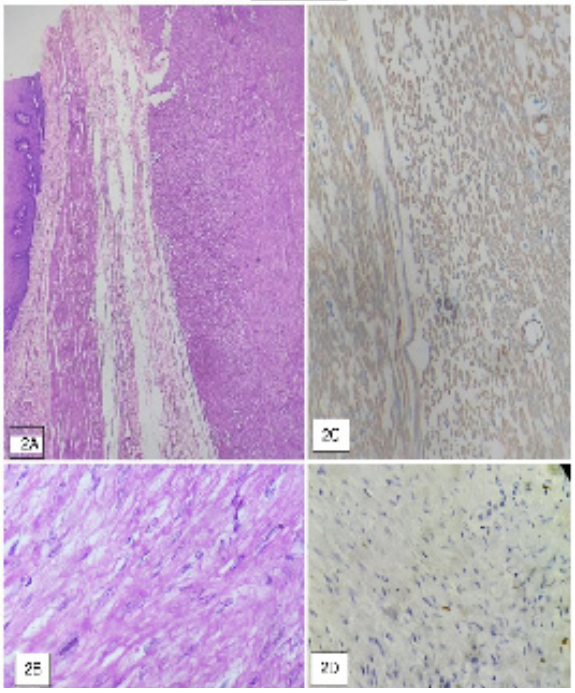


**Figure 2**