

Giant Fibrovascular Polyp – Amending Knowledge About a Rare Entity from a Known Benign to an Unknown Malignant Behavior.

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Abstract

A giant fibrovascular polyp is a non-epithelial tumor of esophagus. It usually arises from the proximal esophagus behind the cricoid cartilage from the upper esophageal sphincter. These polyps were previously considered as benign non neoplastic intraluminal masses. However, with continuous studies and advanced testing by FISH (Fluorescence in situ hybridization) done in multiple cases worldwide, it has now been established that this entity is synonymous with well differentiated liposarcoma. Being proven to be a malignant entity now, care should be taken while treating such cases, as these will require an adequate resection margin during its surgical removal.

This case report aims at enlightening the treating clinicians along with reporting radiologists and pathologists about this rare entity and to emphasize on correct management for patients' benefit as it is known for recurrence and dedifferentiation. Also, being a sarcomatous lesion, it may lead to local and distant spread and prove fatal to the patient.

Keywords: Giant fibrovascular polyp, liposarcoma, esophageal polyp

Introduction

Giant fibrovascular polyps are rare, submucosal indolent intraluminal masses. They usually arise from proximal esophagus. Patient presents with dysphagia, foreign body sensation, regurgitation or sudden asphyxia.^[1]

Diagnosis of these polyps are difficult since clinically and radiologically they mimic benign lesions. Till date there are only a few recent studies which have described this entity as sarcomatous with the help of new markers viz. MDM2 and CDK4.^[2]

Previous studies have described it as a benign lesion obstructing the airway and esophagus.

Case History:

A 63 year old female patient presented with complain of dysphagia for solids and semisolids for 1 month along with prolapse of mass from the mouth occasionally. On esophagoendoscopy, a lesion was identified arising from right pyriform sinus and prolapsing into the esophagus exhibiting bluish discoloration. No other abnormalities seen on examination of oral cavity

or on indirect laryngoscopy. On palpation, no neck nodes were enlarged or palpable.

MRI showed an intraluminal polypoidal and pedunculated growth with smooth margins showing hyperintense signal on T2W/STIR images with no diffusion restriction on DW/ADC images suggestive of benign etiology. It was seen arising from cervical part of esophagus 4.0 cm above lower esophageal junction and was 2.8 cm in diameter.

Treatment plan: Treatment of the patient was planned considering it a benign polyp as per clinical examination and radiological reports. It

was excised from the base and was sent for histopathological examination.

As the lesion was thought to be a benign polyp, a polypoidectomy was considered sufficient as it relieved patient from the symptoms and no further follow up was suggested.

Histopathological examination:

Gross examination showed a large polyp measuring 12 cm in length with a 0.5 cm resection margin. Externally, the surface appeared smooth. On cut section, it was tan colored and soft. No firm areas were identified grossly. Representative sections were submitted.



Figure 1: Gross examination.

On microscopy, fibro-adipose tissue was seen with slightly hyperchromatic stromal cells. Numerous capillary sized blood vessels were seen interspersed amongst adipocytes. Mitotic figures were seen in occasional fields. A diagnosis of giant fibrovascular polyp was made histologically.

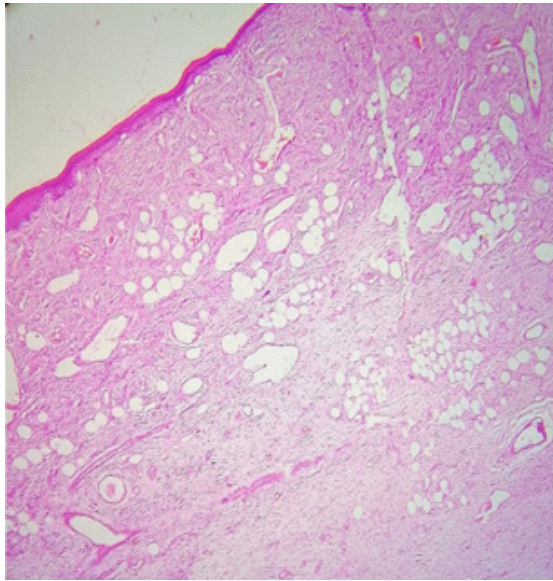


Figure 2. A subepithelial lesion exhibiting areas with nuclear atypia.

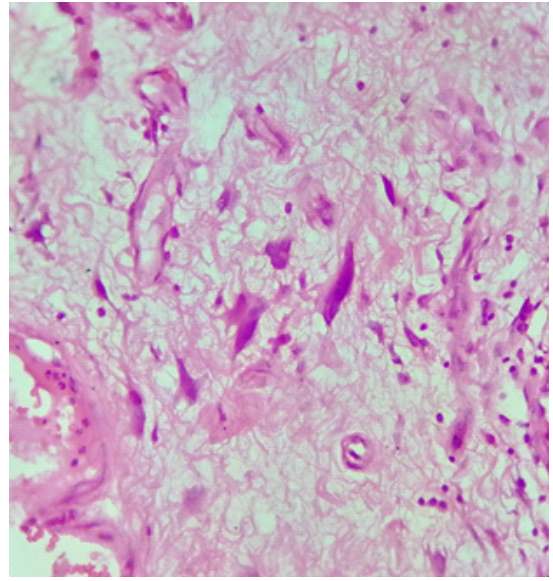


Figure 3. Fibroadipose tissue enmeshed in stromal cells.

However, as it is a very rare entity, IHC was planned for confirmation. On IHC, tumor cells were immunoreactive for p16, CDK4, MDM2, S100 and non-immunoreactive for CD34, CD117, SMA and cyclin D1. Ki67 labelling index was 6-7% in highest proliferating areas. Hence, diagnosis of giant fibrovascular polyp/ well differentiated liposarcoma was confirmed on IHC.

After it was proven to be well differentiated liposarcoma, a PET scan was done to look for residual disease which showed no areas of FDG uptake.

The surgery done had a margin which was free of tumor and the PET scan was negative. Hence, no further

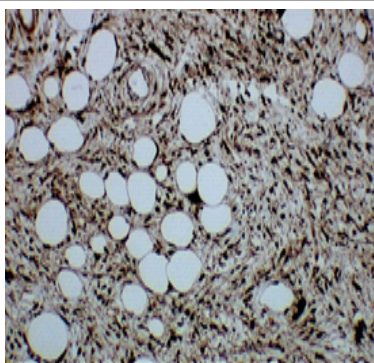


Fig. 4. CDK 4 (EP 180)

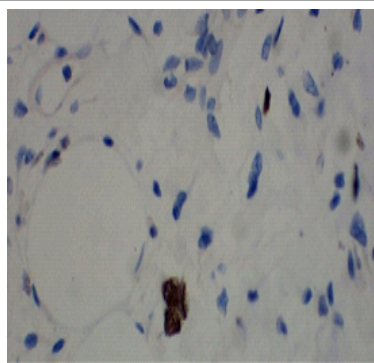


Fig. 5. Ki67 (MIB-1)

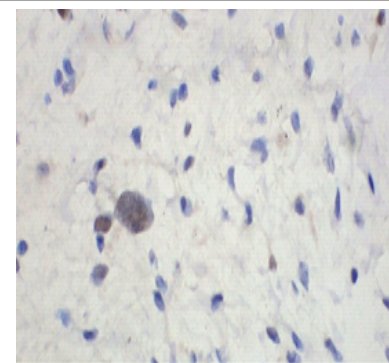


Fig. 6. MDM2 (BSB64)

intervention was required for the patient and she was counselled accordingly and kept on follow up.

Discussion:

Giant fibrovascular polyps of esophagus are rare affecting nearly 0.5% of the adult population.^[3] Out of which, almost 95% of the lesions arise behind the cricoid cartilage as this area is considered weak due to lack of posterior muscular structure with excessive mucosal folds and constant traction from peristaltic activity of esophagus.^[4]

Clinically, patient presents with dysphagia, regurgitation of mass or sudden asphyxia wherein patient will present in emergency. These lesions were traditionally considered benign. However, with more widespread use of immunohistochemical stains, it has become increasingly apparent that these polyps have a malignant behaviour.

There are a very few studies which support malignant behaviour of these lesions. And due to its overlapping features clinically, radiologically and histologically, it can be easily misdiagnosed and patient may land up in recurrence, dedifferentiation or distant spread.

Hence, an in depth knowledge of these lesions is required for all in order to get an accurate diagnosis and to determine patient's management correctly.

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