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Case Report

A Rare Roadblock and An Unyielding Spirit: Inflammatory Myofibroblastic Tumour of the Oesophagus in a Child

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Abstract

Paediatric Inflammatory Myofibroblastic Tumour (IMT) is a rare soft tissue tumour, 1/3rd are reported to be of pulmonary origin and 2/3rd extrapulmonary with oesophagus being a rare site. An early adolescent presented with dysphagia and occasional regurgitation. Endoscopy revealed a friable mass with a wide base at the lower oesophagus; reported as a pyogenic granuloma. Primary resection of tumour with anastomosis to the stomach was performed. Postoperatively child had an anastomotic leak, which was repaired using a diaphragmatic flap. Due to prolonged total parenteral nutrition, he developed bilateral renal and ureteric calculi leading to acute kidney injury causing anuria which was managed by bilateral DJ stenting initially and then stone clearance. Paediatric oesophageal IMT poses a diagnostic challenge due to nonspecific clinical and biopsy findings. Complete resection is the treatment of choice. Diaphragmatic flap could be incorporated as a salvage for oesophageal leaks.

Keywords: Myofibroblastic Tumour; Acute Kidney Injury; Paediatric Oesophageal; Oesophageal Leaks

Introduction

Inflammatory Myofibroblastic Tumour (IMT) is a rare soft tissue tumour classified as a neoplastic disease of intermediate biological potential given the low risk of recurrence and metastatic potential [1]. Number of terms have been used to describe the lesion i.e., inflammatory pseudotumor, fibrous xanthoma, plasma cell granuloma, pseudo sarcoma, lymphoid hamartoma, myxoid hamartoma, inflammatory myofibrohistiocytic proliferation, benign myofibroblatoma and most recently IMT [2]. It is seen amongst children however young adults can be affected [3]. This neoplasm is composed of myofibroblastic mesenchymal spindle cells with an inflammatory infiltrate of plasma cells, lymphocytes and eosinophils [4]. IMT has been reported to occur in multiple locations such as the lung, bladder, spleen, breast, pancreas, liver, colon, spermatic cord, prostate, peripheral nerves and orbit [5]. An exceedingly rare presentation of this tumour is that of in the oesophagus, especially amongst the paediatric population with very few cases reported in published literature. We report a case of an early adolescence male with an IMT of distal oesophagus which was primarily resected with a good outcome.

Case Presentation

An early adolescence boy presented to us with complaints of difficulty in swallowing for 3 months, initially for solids and gradually progressive to involve liquids too. History of occasional regurgitation of food around 3 to 5 minutes after ingestion of food for 1 month was present. There was no history of hematemesis or melena and clinical examination with routine laboratory investigations were unremarkable.

Investigations

An endoscopic evaluation revealed a mass with a wide base in the distal 1/3 of oesophagus and the biopsy was reported as a pyogenic granuloma as depicted on Fig. 1. A contrast enhanced CT revealed an intraluminal growth with mild to moderate luminal narrowing within the distal end of oesophagus measuring 2.8 cm with involvement of gastro oesophageal junction as depicted in Fig. 1.

Differential Diagnosis

Initially a diagnosis of a wide based pyogenic granuloma was considered based on the endoscopic biopsy findings. However, owing to the location of the tumour a malignant adenocarcinoma or a squamous cell carcinoma also was considered as a differential diagnosis. Initially a diagnosis of a wide based pyogenic granuloma was considered based on the endoscopic biopsy findings. However, owing to the location of the tumour a malignant adenocarcinoma or a squamous cell carcinoma also was considered as a differential diagnosis.

Treatment

Initial attempt at laparoscopic mobilisation of the lower oesophagus was abandoned in view of dense adhesions to the crura fearing tumour infiltration. A primary resection of distal end of esophagus with the tumor and the gastroesophageal junction (i.e., 4.5 cm inclusive of tumor and margins) was performed through a right thoracotomy incision and a partial gastric pull up was performed for a tension free gastro -esophageal anastomosis in the lower chest. We additionally performed a Heineke-Mikulicz pyloroplasty and a Witzel feeding jejunostomy. The resected specimen was sent for a histopathological examination as depicted in Fig. 1. Postoperatively from day 2, child was started on total parenteral nutrition and small volume jejunostomy feeds and was extubated on day 4. The biopsy showed spindle cells arranged in fascicle or storiform pattern with intermixed inflammatory infiltrate composed of plasma cells and lymphocytes possibly suggestive of IMT of oesophagus Fig. 2. Immunohistochemistry confirmed the diagnosis of IMT, as the tumour was positive for smooth muscle actin and negative for CD34, CD117, S100 and anaplastic lymphoma kinase.

On post-operative day 7 child developed tachypnoea, tachycardia, fall in saturation with bile-stained output in the intercoastal drainage tube, following which an upper gastrointestinal study was performed revealing an anastomotic leak into the mediastinum as depicted in Fig. 2. Child was reintubated and planned for an emergency thoracotomy as the output through the intercoastal drain was of high output and bilious. Intraoperatively a large rent at the anastomotic site was noted following which the defect was closed and reinforced with a right diaphragmatic flap. The diaphragmatic flap was medially raised as a tongue shaped flap which was sutured to the edges of the anastomotic defect. On post-operative Day 15 children underwent a repeat upper gastrointestinal contrast study revealing no leak with smooth passage of dye into the stomach. Following this the child gradually started on jejunostomy feeds and then oral feeds which he tolerated.

On post operative day 22 while the child was on normal oral feeds, he developed bilateral flank pain with reduced urine output with a rise in serum creatinine of 4.13 mg/dl. In view of this he underwent an ultrasound of abdomen and a plain computed tomography of the renal system which revealed bilateral renal calculi and ureteric calculi, with hydro ureteronephrosis. In view of this he underwent an emergency bilateral ureteroscopy with DJ stenting.

Outcome And Follow-Up

Post operatively his symptoms were relieved and the urine output normalized. He was discharged in good health and is being followed up with resolution of symptoms. He was initially followed up monthly for 6 months documenting weight gain. At 1 year post the primary surgery a contrast study and a surveillance endoscopy were performed which revealed no evidence of recurrence. The child has been on follow with us for 2 years since his primary surgery.

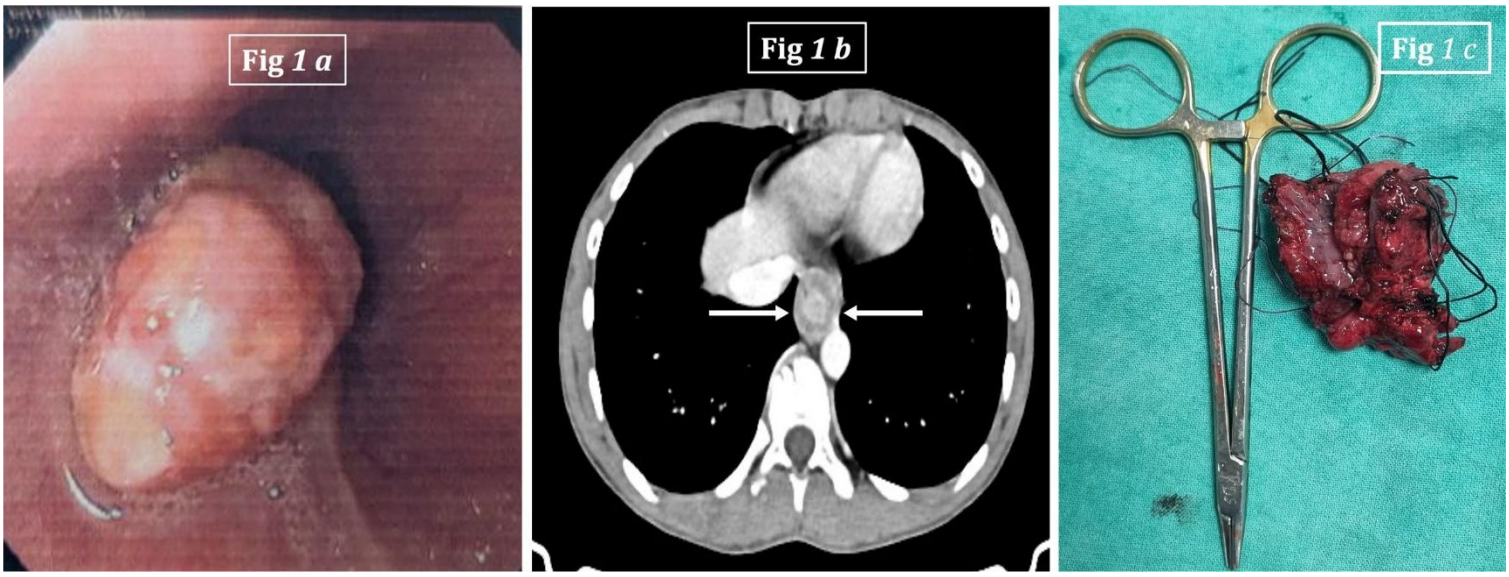

Figure 1: a: Mass at the distal end of oesophagus reported as a pyogenic granuloma; b: Contrast enhanced CT of the thorax depicting an intraluminal growth with mild moderate lumen narrowing within the distal end of oesophagus measuring 2.8 cm with involvement of gastro oesophageal junction; c: Resected specimen of distal oesophagus with the tumour *in-situ*.

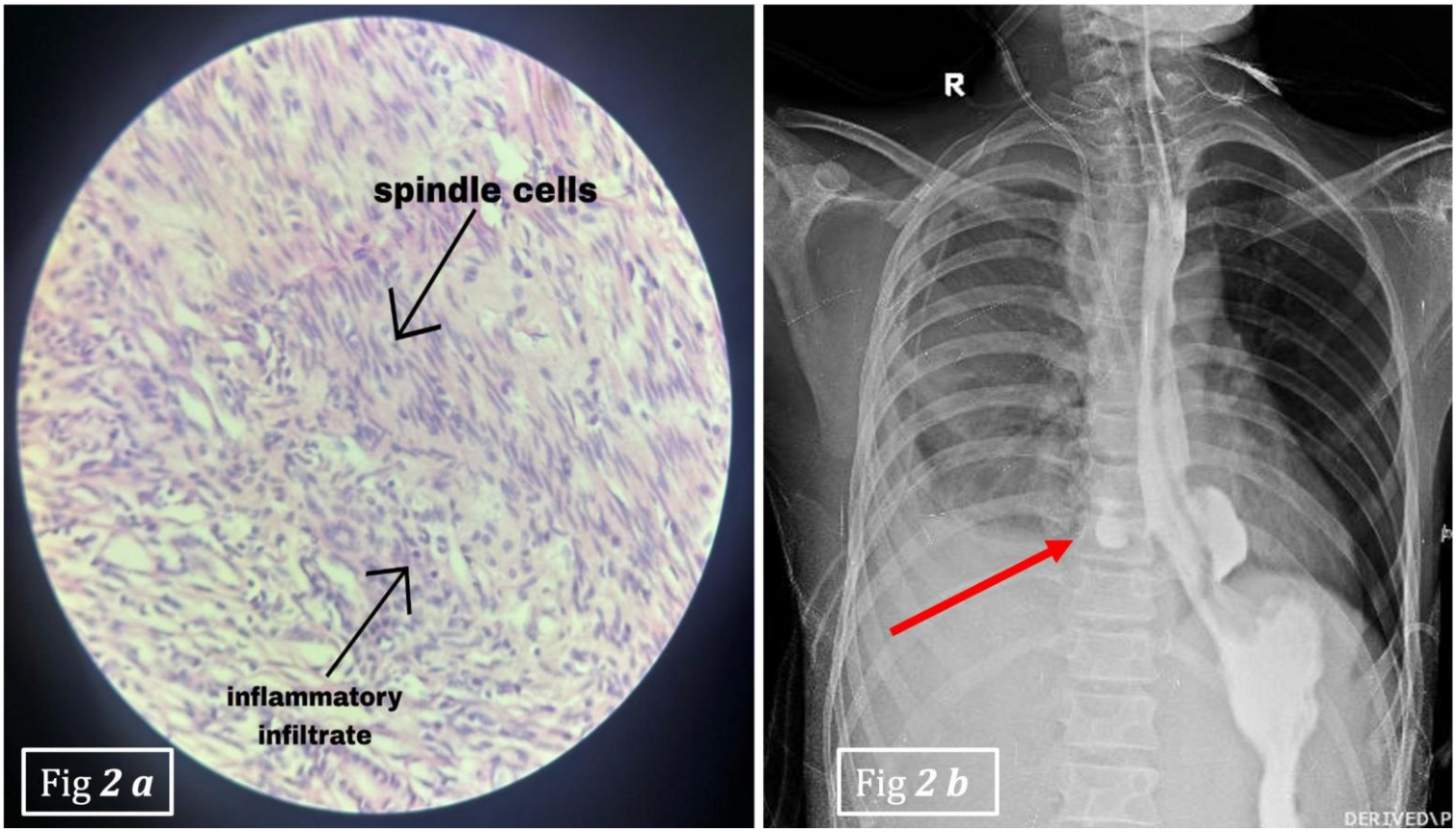

Figure 2: a: Histological slide depicting spindle cells arranged in fascicle or storiform pattern with intermixed inflammatory infiltrate composed of plasma cells and lymphocytes; b: Upper gastro intestinal study depicting the marked area as an anastomotic leak into the mediastinum.

Discussion

Inflammatory myofibroblastic tumour amongst adults, their presentation and management are published in literature, however in children owing to its rarity there are very few published reports. IMT is described as a lesion formed of multiple myofibroblastic spindle cells with associated inflammatory infiltration of lymphocytes, plasma cells and eosinophils and are

regarded as an intermediate-grade tumour that could recur [6]. The exact pathophysiology and aetiology of this tumour have not been fully researched in published in literature. Numerous risk factors, such as smoking, minor trauma, IgG4-related illnesses and aberrant immune reaction to viruses (Epstein-Barr virus and Human Herpesvirus-8) especially amongst adults have been identified [1]. IMT is the most prevalent in the lungs amongst children with oesophagus been an extremely rare site of occurrence [6].

Children with oesophageal IMT often present with progressive dysphagia (as seen in our case), chest pain (usually retrosternal), loss of weight, chronic bleeding with iron deficiency anaemia and elevated temperature (caused by cytokine release) [7]. Literature indicates challenges in diagnosing (endoscopic and radiological) oesophageal IMT, with most tumours identified post-resection [8]. In the above case the child underwent 2 upper gastrointestinal endoscopies with biopsies and both were in conclusive of the diagnosis and was provisionally diagnosed as a case of pyogenic granuloma of the oesophagus. The preferred diagnostic procedure is an ultrasound guided endoscopy which reveals a hypoechoic lesion of the muscularis propria. However, in just 77% of cases, there is a correlation between endoscopic ultrasound characterisation and ultimate pathology [7]. On gross examination IMT s are solid tumours made up of myofibroblastic cells in the shape of spindles that have plasmacytic infiltration. The cells are immunopositive for Vimentin (77%) and smooth muscle actin (86%) [7]. Only 50–70% of tumours have ALK gene rearrangement, even though it is thought to be a crucial molecule for diagnosing IMT s and if ALK is positive it represents higher rates of malignancy and increased rates of recurrence [3]. In our case report smooth muscle actin was reported to be positive and negative for CD34, CD117, S100 and anaplastic lymphoma kinase which helped us to confirm the diagnosis of IMT.

Radical surgery is the principal treatment modality in IMT s of the esophagus which entails an esophagectomy in the case of oesophageal location. The primary method of oesophageal replacement is gastric transposition. It is reported that the preferred treatment for big (>2.5 cm) or obstructive oesophageal IMTs should be an oesophageal resection, whether partial or complete. Similarly, in our case we performed complete resection of the lower end of the oesophagus with the tumour, with a gastric pull up to facilitate gastro oesophageal anastomosis. We performed an additional pyloroplasty to facilitate easy flow off gastric contents with a feeding jejunostomy to start early enteral feeds.Two cases of successful endoscopic resection of IMT have been reported, of which the 1st underwent a piecemeal endoscopic mucosal resection with a hot snare, while the 2nd underwent successful endoscopic resection with no signs of tumour recurrence at the annual follow-up [9]. Endoscopic resection is successfully possible only in case of smaller and pedunculated narrow base tumours. Despite IMT's reported to be a benign tumour, poly nodularity, irregular margins and unclear borders are risk factors for recurrence and metastasis.

Both early and late complications following a esophagectomy are documented (59%), commonest been an anastomotic leak (11.4 %) [10]. Smaller leaks do settle with a trial of conservative management, but larger leaks require immediate re-exploration. We in our case, attribute the cause of the anastomotic leak to be secondary to the discrepancy between the distal esophagus and the proximal end of the stomach causing ischemia. As reported in literature, intrathoracic esophageal leaks associated with systemic sepsis require prompt surgical exploration, debridement of all necrotic and devitalized tissue with a secure closure of the leak [11]. We similarly encountered an anastomotic site leak in our child following which initially, he was conservatively managed which failed as he developed signs of sepsis which necessitated an immediate exploration. We incorporated the usage of the right diaphragmatic muscle flap as a cover over the anastomosis which is similar to that reported by Richardson, et al., where in a diaphragmatic muscle flap was used without attempting a primary closure of the defect because of tissue friability [12]. Use of other salvage flaps such as intercoastal muscle flaps and pleural wraps have also been reported in literature. A repeat contrast study in our child revealed no evidence of leak with a smooth flow of contrast across the anastomotic site.

Conclusion

It is well-established that prolonged usage of total parenteral nutrition can cause complications such as infections from central venous catheter, liver damage (i.e. fatty liver, cholestasis, cirrhosis), metabolic issues (i.e. hyperglycaemia, electrolyte imbalances, micronutrient deficiencies), thrombosis of veins and bone diseases like osteoporosis [13]. A much rarer complication is hypercalciuria leading to the formation of renal calculi which we encountered in our case [14]. In our case the child developed acute kidney injury secondary to bilateral obstructive renal and ureteric calculi which caused complete obstruction to the renal system, for which he required an emergency bilateral DJ stenting, following which the acute kidney injury resolved. At follow up the child has gained weight with no recurrence of symptoms.

Even though the child underwent multiple procedures i.e., primary resection of the tumor, re exploration for anastomotic leak and bilateral DJ stenting; the unyielding spirit of the child helped us to manage and tide over all these morbid complications.

Oesophageal IMT in children is a diagnostic rarity that often eludes preoperative confirmation due to non-specific biopsy findings. A complete surgical resection remains the cornerstone of treatment, offering favourable prognosis when combined with meticulous post-operative care.

A diaphragmatic flap can be used as salvage flap in case of oesophageal leaks with a good outcome. Awareness about this tumour, coupled with high clinical suspicion with a multidisciplinary approach, is crucial in reducing the morbidity and mortality amongst children.

Learning Points

- Oesophageal inflammatory myofibroblastic tumour in children is a diagnostic rarity that often eludes preoperative confirmation due to non-specific biopsy findings
- A complete surgical resection remains the cornerstone of treatment
- A diaphragmatic flap can be used as salvage flap in case of oesophageal leaks
- Morbidity associated with prolonged total parenteral nutrition should be monitored

Conflict of Interest

The authors declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

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Data Availability Statement

The data supporting the findings of this study are available from the corresponding author upon reasonable request.

Ethical Statement

The project did not meet the definition of human subject research under the purview of the IRB according to federal regulations and therefore was exempt.

Informed Consent Statement

Informed consent was obtained from all participants included in the study.

Authors' Contributions

The following authors were responsible for drafting of the text, sourcing and editing clinical images, investigation results, drawing original diagrams and algorithms and critical revision for important intellectual content: VV and NP. The following authors gave final approval of the manuscript: VV, NP, PSK and AS. PSK is responsible for the overall content as guarantor.

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