Pedunculated, well differentiated liposarcoma of the oesophagus mimicking giant fibrovascular polyp

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ABSTRACT

We present a rare case of a big oesophageal liposarcoma causing dysphagia and weight loss in a 75-year-old patient. Endoscopically, a pedunculated lesion with subtotal obstruction of the oesophageal lumen had been detected and thoracoabdominal oesophageal resection with gastric sleeve reconstruction was performed. Surprisingly, a liposarcoma of the oesophagus was revealed on histopathological analysis, showing *MDM2* overexpression. Oncological follow-up has been uneventful and the patient remains in good clinical shape at 15 months after surgery.

KEYWORDS

Giant fibrovascular polyp – Well differentiated liposarcoma – Thoracoabdominal oesophageal resection – *MDM2* overexpression

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Case history

A 75-year-old patient presented with marked dysphagia leading to unwanted weight loss necessitating total parenteral nutrition. Endoscopically, a near-to-total obstruction of the oesophagus could be detected (between 18cm and 39cm from the incisors) caused by a mass of soft, jelly-like consistency originating from the oesophageal wall (Fig 1). Furthermore, there was long segment Barrett's oesophagus, an axial hiatal hernia and minimal antral gastritis. After the initial workup, the patient was transferred to our upper gastrointestinal centre for further diagnosis and treatment.

Endoscopic ultrasonography revealed a semicircumferential spongiform ballooning of the submucosal tissue of the oesophageal wall up to 6.5cm in its longest diameter from 18cm to 43cm from the incisors showing a heterogeneous structure of the lesion with fewer echogenic areas in the distal part. Neither infiltration of the muscularis propria nor signs of lymphadenopathy could be detected (Fig 2). Computed tomography (CT) showed a stenosis of considerable length with pemphigoid-like ballooning of the whole oesophagus, indicating possible malignancy (Fig 3). Our multidisciplinary gastrointestinal tumour team recommended resection of the stenotic part of the oesophagus.

An oesophagectomy was performed, starting with the thoracotomy in order to gain more information on the extent of the tumour. On exploration, tumour growth proved to be confined to the oesophageal lumen without infiltration of the surrounding area. An additional cervical incision was

made to enable complete removal of the lesion (Fig 4). Tension free reconstruction was achieved with a long gastric sleeve being formed after abdominal exploration and two-field lymph node dissection (Fig 5).

The postoperative course was uneventful. The patient could be weaned quickly from mechanical ventilation and oral nutrition was started on the first postoperative day as

Figure 1 Endoscopy showing the stenosing mass originating from the oesophagus (star)

Figure 2 Endoscopic ultrasonography showing ballooning of the muscularis mucosa (arrow)

Figure 4 Resected specimen showing the oesophagus (arrow) and pedunculated polyp (star)

Figure 3 Computed tomography of the oesophageal mass (star)

Figure 5 Gastric sleeve before suturing of the anastomosis (arrow)

per the hospital's routine protocol. An endoscopy was performed on day 2, showing a vital anastomosis. A postoperative barium swallow confirmed correct passage without evidence of stenosis or anastomotic insufficiency. Seventeen days after surgery, the patient was discharged from hospital.

Clinical follow-up review at four and ten months and fifteen after surgery found the patient in good overall condition, and he was regaining his preoperative weight. Endoscopy findings were normal and there was no sign of tumour recurrence.

Histopathological findings

The resected tumour specimen measured 25cm in length and 10cm in diameter. The growth was mainly intraluminal and originated from the left oesophageal wall. Its consistency was gelatinous. There were multiple ulcerations of the overlying squamous mucosa. Histopathologically, the tumour consisted of fibrotic and fatty tissue, and was highly vascularised without obvious signs of neoplasia (Fig 6). Immunohistochemical analysis revealed proliferative activity as indicated by Ki-67 positivity in the basal layers of the squamous epithelium and in the germinal centres of the lymphoid follicles whereas the lesion itself had a Ki-67 index of 2%. CD117 expression could be demonstrated only in

mast cells while CD34 positivity was observed in the vascular plexus as well as in some spindle cell areas. Fatty tissue was positive for S100 protein and perivascular smooth muscle cells showed positivity for smooth muscle actin.

Based on these findings, the initial differential diagnoses included GFP and well differentiated liposarcoma (WDL). Further histological analysis revealed very few atypical spindle cells and occasional lipoblasts. Finally, fluorescence in situ hybridisation detected cluster amplification of the *MDM2* gene, ultimately determining the diagnosis of a partly sclerotic WDL with inflammatory parts (Fig 7). No lymph node infiltration was noted.

Discussion

The differential diagnoses of pedunculated, intraoesophageal masses include giant fibrovascular polyps (GFPs) as well as well-differentiated liposarcoma (WDLs), which are much rarer. Only about 100 cases of GFP have been described so far.^{2,5} The largest series, reported by Levine *et al*, consists of 16 polyps.⁴ GFPs account for approximately 2% of benign oesophageal tumours.⁵ They typically arise in middle aged patients, with a male-to-female ratio of 3:1.^{1,5}

Figure 6 Highly differentiated adipose tissue (arrow)

Figure 7 MDM2 gene amplification (arrow)

Single cases of GFP in children have been reported as well.⁶ Patients with GFP mainly present with clinical signs of dysphagia (87%), dyspnoea (25%), regurgitation of the growing polyp (12%), weight loss or anaemia due to occult bleeding.^{2,5,7} In some cases, regurgitation of the polyp can compromise the airways. Around a third of patients die owing to an oesophagus polyp causing respiratory distress, with symptoms starting to occur when the polyp grows to a size of about 5cm in length.^{1,8}

Polyps generally originate from redundant mucosal folds in Laimer's triangle in the cervical section of the oesophagus. The aetiology remains unclear but genetics, prior trauma and irradiation play a role in the pathogenesis.^{3,8} Polyps usually grow aborally from their point of origin, manifesting as long intraluminal masses, which can reach into

the stomach in extreme cases.⁹ This is thought to be because of the direction of the swallowing act.¹⁰

The extent and localisation of the mass define the therapeutic options. The preinterventional diagnostic workup should include barium studies, upper gastrointestinal endoscopy including endoscopic ultrasonography and CT or magnetic resonance imaging (MRI) for staging purposes.⁵ Oesophagogastroduodenoscopy with consecutive application of a contrast agent helps to determine the size and relative position of the tumour and the extent of oesophageal stenosis as well as any suspicious mucosal lesion, which should always be biopsied.

As polyps are often covered with normal mucosa and the oesophagus can dilate if there is stenosis or air insufflation, polyps may be missed on endoscopic evaluation.^{1,8} Endoscopic ultrasonography is required to further characterise the tumour and determine the tumour's origin by finding the peduncle; it can also determine vascularisation as well as depth invasion and surrounding lymphadenopathy.^{1,5} On endoscopic ultrasonography, GFPs appear as smooth, ballooning, often sausage-like masses, as in our patient.^{5,9} Barium swallow provides a functional view of the mass and helps to distinguish it from fixed stenosis, achalasia or other causes of oesophageal obstruction.^{1,2} CT or MRI studies are mandatory and can detect local infiltration or distant metastasis.^{1,5,7,9,10}

Oesophageal polyps should be resected completely to prevent recurrence, malignant transformation and further complications such as regurgitation, asphyxia or death.^{1,7,11} Resection can be performed endoscopically (if the polyp is not too big) or surgically. In selected cases, laser ablation has been reported to result in less trauma to the surrounding tissues, lower morbidity and a shorter hospital stay than more invasive procedures.^{1,5,7}

Endoscopic removal of the polyp with curative intent is only applicable to smaller, mobile polyps with a small base. With the help of electric loops or rat bite graspers, the polyps can be removed in a manner similar to colonic polyp removal.^{5,8} If hypervascularisation is diagnosed, advanced haemostatic techniques should be available.⁹ Tracheal intubation helps to create a safe working environment.⁵

If complete removal of the mass cannot be achieved endoscopically, surgery is required.¹ Surgical technique should be chosen based on the location and size of the polyp.⁹ Smaller polyps in the proximal oesophagus can be removed via a cervical oesophagotomy or pharyngotomy. In these instances, the exact localisation of the peduncle should be known beforehand so that the procedure can be started from the opposite side.⁷ For bigger masses or those located atypically, a transthoracic oesophagectomy through a right thoracotomy or a combined thoracoabdominal procedure may be necessary.⁷ If the patient is unable to tolerate intermittent single lung ventilation or if the tumour is located distally, a transhiatal oesophagectomy without thoracotomy may be possible.

Regardless of the procedure chosen, reconstruction should be performed constructing a gastric sleeve.¹² If this is not possible, then left or right colon, jejunum or free

revascularised grafts may be used to reconstruct the defect. In cases of suspected malignancy, a two-field lymphadenectomy should be performed as well.

Depending on the type of procedure carried out, the patient should be transferred to the intensive or intermediate care unit following surgery. Early postoperative extubation is recommended. Postoperative nutrition should be started slowly and with care.

On histopathological examination, GFPs are usually composed of adipocytes, vascular epithelial structures and fibrous tissue covered with mature squamous mucosa; this can lead to a GFP being mistaken for a fibroma, fibrolipoma, fibromyxoma or lipoma.¹¹ The World Health Organization includes all of these under the umbrella term 'fibrovascular polyp'.¹¹ However, WDL of the oesophagus should also be considered as a differential diagnosis in such cases.¹²

WDLs are the second most frequent type of liposarcomas, often located in soft tissue of the limbs or the retroperitoneum.^{5,15} Liposarcomas of the gastrointestinal tract are rare.^{12,15} As the number of atypical cells and lipoblasts may be low in WDL, the final diagnosis often only becomes apparent after molecular analysis.¹⁵ For WDLs, giant marker ring chromosomes with *MDM2* amplification can usually be detected, as in our patient.^{12,15} In up to 90% of cases, there is additional *CDK4* amplification as the gene is located in close vicinity to the *MDM2* gene.^{12,15} In order to rule out other potential differential diagnoses such as gastrointestinal stromal tumour, schwannoma or inflammatory fibroid polyp, additional immunohistochemical staining for DOG1, S100 and desmin can be helpful.¹²

If a WDL of the oesophagus is found on histopathological analysis, presentation of the case in a multidisciplinary tumour team meeting and oncological follow-up review are mandatory. While the risk for local recurrence can be up to 80% for WDL of the oesophagus,⁵ if incomplete resection is performed, lymph node infiltration is rare. Nevertheless, recurrence in WDL has been described after a relatively long interval.⁵ As a result of the high recurrence rate and the possibility of delayed recurrence, some clinicians favour radiotherapy after incomplete resection. On the other hand, distant metastasis of GFP or WDL has never been reported.^{1,5} If the tumour can be removed completely, adjuvant treatment is not necessary.

Conclusions

Only few cases of GFP have been reported in the literature and these have been mainly described as benign processes of the hypopharynx or oesophagus.^{5,10} Owing to their size,

they may nonetheless cause severe or even life threatening complications and should therefore be resected.

Modern techniques can reveal gene mutations, which can help when making the diagnosis of WDL of the oesophagus. Irrespective of whether malignancy is suspected, the preoperative workup (including exact staging via endoscopy and endoscopic ultrasonography, with biopsies and CT/MRI) should be the same for both types of polyp.⁵ Therapeutic approaches include endoscopic or surgical removal. If it is uncertain whether the growth is benign or malignant, radical oncological resection is usually advised by a mandatory preoperative multidisciplinary tumour team. Complete removal of the mass is required to prevent recurrence. New histopathological and molecular techniques help to achieve a more detailed characterisation of the resected tissues. Liposarcoma of the oesophagus should always be excluded by employing immunohistochemistry and molecular analysis to exclude aberrations in *MDM2* and *CDK4*.¹⁵

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