Rare Case of Primary Diaphragmatic Leiomyoma

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Abstract

Diaphragmatic tumors are rare pathology. Little over 200 cases are reported in literature with histological variance being very diverse. The incidence of malignant and benign diaphragmatic tumors is relatively similar, with most common being cysts and lipomas in benign group versus sarcomas in malignant group. Due to tumor anatomical localization many patients do not have any specific symptoms until tumor size increases. Right diaphragm tumors can mimic liver lesions – solid as well as cystic. Surgical resection is the only curative option. This case report aims to provide clinical experience and diagnostic challenges of one of the rarest benign diaphragmatic tumors – leiomyoma.

Keywords: Diaphragm; Diaphragmatic tumor; Diaphragmatic leiomyoma

Introduction

Diaphragmatic tumors are rare entity. Due to rarity no systematic analysis is possible and the researches on these tumors are limited to case reports and case series. Little over 200 cases are reported in literature with histological variance being very diverse [1]. The incidence of malignant and benign diaphragmatic tumors is relatively similar, with most common being cysts and lipomas in benign group versus sarcomas in malignant group [2]. Because of the rarity most of the tumors are diagnosed relatively late and have latent clinical presentation. It also presents a diagnostic challenge in differentiating diaphragmatic lesions from liver lesions or from other organ tumors [3]. The extent of surgical resection and reconstruction is also controversial, since the radical resection may lead in need of diaphragmatic prosthesis [4]. This case report aims to provide clinical experience and diagnostic challenges of one of the rarest benign diaphragmatic tumors – leiomyoma.

Case Presentation

A 48-year-old patient received left laparoscopic oophorectomy due to serosal proliferative tumor in gynecological department. This procedure was followed by right laparoscopic oophorectomy, omentectomy and biopsies with negative histological findings. In postoperative period patient started complaining with abdominal distress. Ultrasound was performed, which revealed 8.3 cm × 14 cm solid mass in liver S6/7 segment Figure 1. Abdominal MRI was recommended for differential diagnosis during follow-up period. Following tumor markers – alpha fetoprotein, carcinoembryogenic antigen and Ca 19.9 were in normal range. Abdominal MRI revealed 95 mm × 120 mm × 96 mm solid mass in liver S7/8 segment and right sub diaphragmatic space with cystic degeneration Figure 2. Multidisciplinary team, including multiple surgeons, radiologist, thoracic surgeons, and oncologists, reviewed clinical case and Magnetic Resonance Imaging (MRI) images prior to hospitalization and surgery and consensus was made that tumor was not of liver but diaphragmatic origin. Patient was prepared for surgery. Right subcostal incision was made. Following intraoperative examination confirmed that tumor was originating from right diaphragm and pressing on to liver S7/8 segment. The tumor was mobilized by blunt dissection from liver surface followed by mobilization of right liver lobe. Right diaphragm was exposed. The base of the tumor together with surrounding diaphragm was resected using ultrasound knife. The defect in diaphragm was repaired with continuous prolene suture Figure 3. Postoperative period was uneventful. Patient was discharged at eighth post-operative day. Pathological examination showed grayish in capsulated solid 17.5 cm × 10.5 cm × 7 cm tumor with positive expression of estrogen and progesterone reaction. Following protein expression was investigated: SMA: 100% (+++), Desmin:...
100% (+++), H-caldesmon: 100% (+++), Estrogen receptors: 90% positive tumor cells (+/+++), Progesterone receptors: 20% positive tumor cells (+/+++), Ki67: 1% of positive tumor cells (+/+++). HMB45, Melan A, Myogenin, MyoD1, DOG1 – was negative Figure 4.

Discussion

Leiomyomas are generally benign smooth muscle tumors with very low malignancy potential, which is around 0.1% [5]. Most common place of origin of leiomyomas is uterus followed by gastrointestinal tract. Clinicopathological presentation usually depends on place of origin and is often occult, resulting in clinical manifestation when tumor grows to extreme sizes. Primary diaphragmatic leiomyoma is extremely rare, to our knowledge only one published case report exists [6]. One of the common pitfalls in interpretation of diaphragmatic tumors is related to anatomical
position. Tumors of right diaphragm can often be mistaken with liver lesions, especially if diaphragmatic cysts are considered [3]. Most common radiological method for differential diagnosis is abdominal computed tomography or magnetic resonance imaging [7,8]. In our case, MRI was used to specify origin of lesion, however only during multidisciplinary team meeting correct diagnosis was achieved. Thus, emphasizing the necessity of multiple evaluations of imaging. In the era of minimally invasive surgery, there are several diaphragmatic resection techniques to consider. Depending on the tumor involvement and differentiation, partial or complete resection of diaphragm may be needed, following simple suture closure or prosthetic mesh reconstruction [4]. Open or minimally invasive approach can be considered. The main aspect to consider choosing surgical approach is tumor size, integrity, cystic vs. solid, and most important - the radicality of resection, because surgical resection is the only curative option [9]. Minimally invasive approach is usually better suited for patients with small lesions, whereas with big tumors may require extensive liver mobilization or retroperitoneal dissection [10,11]. Since benign tumors usually do not involve large area of diaphragm resulting in smaller resection defect, simple closure with interrupted or continuous suture is sufficient, as was done in our case, where diaphragmatic involvement was around 2 cm in diameter and was closed with unobservable continuous suture [12]. Several case reports have reported that the higher expression of Ki67 is associated with malignancy potential in benign diaphragmatic tumors, whereas in our case, Ki67 expression was positive in 1% of tumor cells [13,14]. Recurrence rate and survival rate varies greatly and depends on tumor histological type, especially malignant ones, and can range from 54% to 89% in long term survival [15,16].

In conclusion, diaphragmatic tumors pose a diagnostic, clinical and surgical challenge due to their latent clinical presentation and anatomical localization, since the resection is the only curative way. Our experience showed that multiple evaluations of radiological images in multidisciplinary team is paramount in choosing correct treatment modality.

References

