GENERAL UROLOGY
Case Report

Benign diaphragmatic neurilemmoma mimicking a left adrenal cyst

Sol adrenal kisti taklit eden benign diyafragma nörilemomu

Rohit Bhattar, Vinay Tomar, Devendra Singh Dhakad, Neeraj Agarwal

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ABSTRACT

Neurilemmomas are benign, slow growing, encapsulated nerve sheath tumor. These tumors arise from the schwann cells of neural crest. Neurilemmomas can manifest in various form according to site, extent and severity of involvement of organ. Diaphragmatic neurilemmomas are very unusual and even difficult to diagnose on preoperative imaging. We will report a case of 39 year old male, who presented with complaints of occasional left flank pain for one year and subsequently investigated, which showed left adrenal cyst with haemorrhagic fluid content. On the contrary, when surgical exploration of the lesion was done, it showed a cyst within the diaphragm, completely separated from left kidney and left adrenal with haemorrhagic content in situ. Histopathological examination of the lesion showed it to be a benign neurilemmoma with cystic degeneration. Post opearatively patient did well. We believe that this is the first case report, which is addressing such kind of initial manifestation of diaphragmatic neurilemmomas.

Keywords: Adrenal; benign; diaphragmatic; neurilemmoma..

ÖZ

Nörilemomlar iyi huylu, yavaş büyüyen, kapsüllü sinir kılıfı tümörüdür. Bu tümörler nöral kristanın schwann hücrelerinden çıkar. Nörilemomlar organ tutulumunun yeri, yaygınlığı ve şiddet derecesine göre farklı şekillerde ortaya çıkabilir. Diyafragmatik nörilemomlar çok nadir olup hatta ameliyat öncesi görüntülemelerde bile tanı koymak zorlaşır. Otuz dokuz yaşında bir yıl boyunca ara sıra gelen sol yan ağrısı yakınmaları olan ve daha sonra araştırıldığında kanlı sıvı içerikli sol adrenal kiste show gösterilen bir olguyu raporluyoruz. Aksine lezyonun cerrahi açınımı yapıldığında diyafram içinde sol böbrek ve sol adrenalden tamamen ayrı içeriği kanlı bir kist olduğu gösterilmiştir. Lezyonun histopatolojik incelemesi kistik dejenerasyona uğramış iyi huylu nörilemomu göstermiştir. Hastanın postoperatif dönemi iyi geçmiştir. Bu olgu raporunun ilk kez diyafragmatik nörilemomun başlangıç belirtilerini ele alan ilk bildirim olduğuna inanıyoruz.

Anahtar Kelimeler: Adrenal; benign; diyafragmatik; nörilemom.

ORCID IDs of the authors:
R.B. 0000-0002-2235-1317;
V.T. 0000-0002-3517-7231;
D.S.D. 0000-0003-4696-8060;
N.A. 0000-0001-8105-346X

Department of Urology, Sawai Man Singh Medical College, Jaipur, Rajasthan

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Correspondence:
Rohit Bhattar
E-mail:
bhattarrohit@gmail.com

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Introduction

Neurilemmomas are unusual tumors arising from Schwann cells of the peripheral nerve sheath. They are also referred as schwannomas. In terms of their morphology and clinical characterstics, these tumors are among the most varied of human neoplasm and are subjected to frequent misdiagnosis.[1] Primary diaphragmatic neurilemmoma is an extremely rare tumor and usually benign in nature.[2] Although schwannomas are usually found in the head and neck region, they have been reported in the gastrointestinal tract in which stomach and small intestine are the most common sites and rarely liver, pancreas, kidney, brain, heart, adrenal, retroperitoneum and diaphragm can be involved.[1-7] They can also present as an unusual manifestation of an infrequent lesion as seen in this report. The diagnosis is mainly one of the exclusion and usually based on histology. Usually these tumors are diagnosed incidentally on evaluation of symptoms like in this case, in which patient's only symptom was

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occasional left flank pain. So these patients are often initially misdiagnosed and final diagnosis is made only after biopsy report. The aim of our case report is, to spread awareness about the occurrence of diaphragmatic neurilemmomas as an unusual presentation, so that prompt treatment could be individualized according to the site, extent and severity of involvement.

Case presentation

A 39 year old, otherwise healthy male, was presented in our department with complaints of occasional dull aching left flank pain for 1 month. General physical examination was unremarkable. He denied any symptoms of headache, palpitations, anxiety, decreased appetite, weight loss or any urinary symptoms. Routine blood investigations were within the normal range. Ultrasound abdomen suggested a 7x7 cm left adrenal mass with internal echoes and solid nodule at its lateral wall. Magnetic resonance imaging (MRI) abdomen showed a 10x6 cm cystic lesion in left suprarenal lesion. Lesion was hypo intense on T1 weighted images and hyper intense on T2 weighted images that showed mild enhancement on intravenous administration of gadolinium contrast (Figure 1a, b). Routine hormonal evaluation for adrenal gland like serum cortisol, 24 hour urinary metanephrines were within the normal range. Presumptive diagnosis of left adrenal complex cyst was made. So surgical exploration was planned and around 10x6 cm well circumscribed cystic structure was identified within the diaphragm which was completely separated from the left adrenal, left kidney and remaining surrounding structures (Figure 2a). Tumor was excised in toto. Content of this tumor was hemorrhagic. Amylase and lipase of this fluid was within normal range. Gross appearance showed well defined cystic structure with areas of degeneration (Figure 2b). Tumor was sent for histopathological examination. Microscopy showed verocay bodies (Figure 3a), cystic changes (Figure 3b) and hyper and hypo cellular areas called as Antony A and Antony B respectively (Figure 3c). No significant mitotic activity or cellular atypia were noted. Final impression was benign diaphragmatic schwannoma with cystic degeneration and normal left adrenal

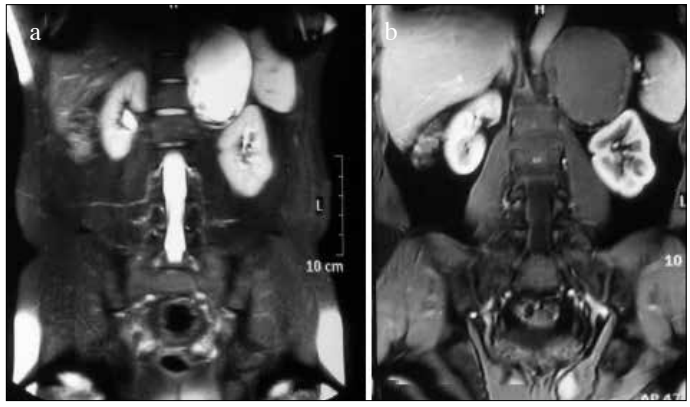

Figure 1. a, b. (a) T2 weighted MRI image showing left adrenal cyst. (b) After gadolinium contrast administration cyst showing mild enhancement

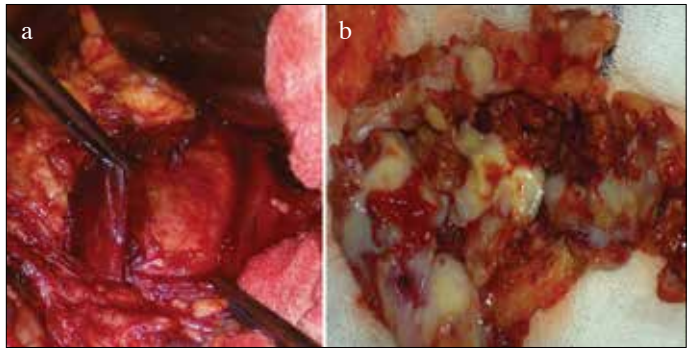

Figure 2. a, b. (a) Intraoperative photograph showing tumor within the diaphragm (straight arrow showing diaphragmatic leaflet and curved arrow showing cyst). (b) Gross appearance of excised tumor

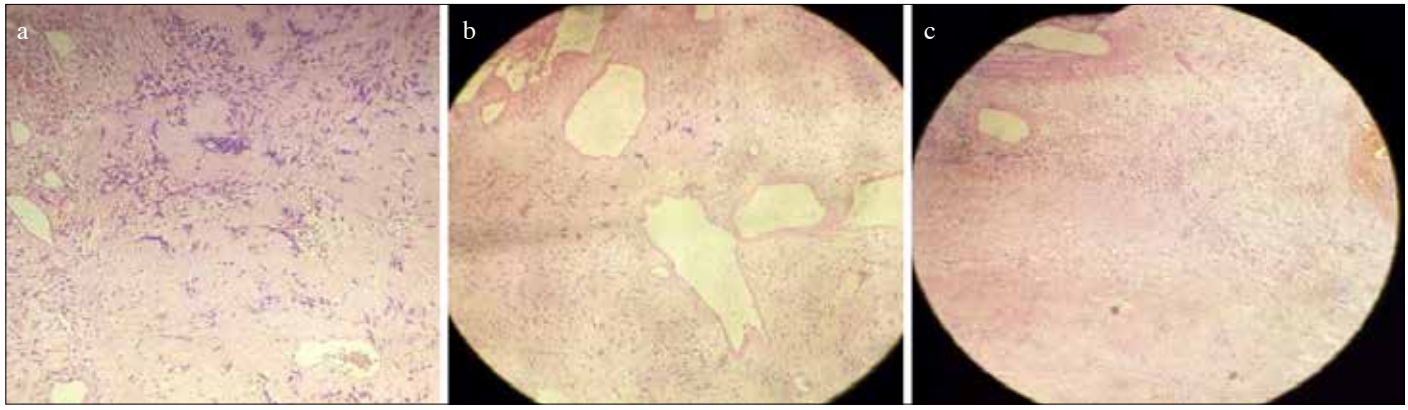

Figure 3. a-c. (a) Verocay bodies (40 x magnification, H & E staining) (arrow showing verocay bodies). (b) Cystic spaces (10 x magnification, H & E staining) (arrow showing cystic spaces). (c) Hypercellular or Antony A area (straight arrow) and Hypocellular or Antony B area (curved arrow) (10 x magnification, H & E staining)

gland. Postoperatively patient did well and repeat imaging done (Contrast enhanced computed tomography) which showed that there is no remaining lesion and both adrenal glands were normal.

Discussion

Neurilemmomas are slow growing, encapsulated nerve sheath tumor that are mostly benign in nature. These tumors usually seen in the adults between the ages of 20 and 50 but can be seen in any age group including children.[8] Patients are generally asymptomatic and diagnosis may be misleading on routine investigations. Diaphragmatic neurilemmomas are very rare and represent approximately 3% of all diaphragmatic neoplasm thus, only very few cases have been reported.[9] These tumors are usually not infiltrative and symptoms occur due to mass effect. Diagnosis is often difficult in such unusual location and usually tumor increases large enough to produce symptoms. Most common presenting symptom is dull aching abdominal pain. Usually site of origin for these tumors are cranial and peripheral nerves. Although benign in nature, in rare cases it may transform into malignant degeneration, having high potential for local recurrence and distant metastasis. The diagnosis of malignant tumor lacks standardized diagnostic criteria but features dense fascicles in a "marble like" pattern consisting of asymmetrically tapered spindle cells.[10] In contrast to benign schwannomas, malignant ones having high recurrence rate and poor prognosis. MRI is the imaging modality of choice for most soft tissue lesion; however in our case MRI was misleading.[11] Few case reports had suggested that it can present as an adrenal mass, either arising from adrenal or as a diagnostic dilemma.[7] Similarly in our case initially it was suspected a case of left adrenal cyst but surgical exploration and subsequent final histological examination confirmed it as a benign schwannoma with cystic degeneration and completely free adrenal gland. To the best of our knowledge, only 13 cases have been reported in the literature of diaphragmatic neurilemmomas (Table 1).[2-15] However present case is unique one, as this is the first case having such kind of dilemma in its initial presentation, where initially there was no clue regarding involvement of diaphragm but during surgical exploration and with final histopathological examination, whole scenario was changed. No other previous report had showed such kind of initial presentation. As in some previous studies, our patient also had schwannoma localized within the diaphragm with no involvement of any other organ and confusion occurs with imaging studies.[12-15] In our case, even though mass was having a big size, during surgery, we found that it was cystic and completely free from surrounding structures. Schwannoma can also present as very usual manifestations like in the form of diaphragmatic schwannoma mimicking hydatid cyst, liver tumor, hypertrophic pulmonary osteoarthropathy, gastric mass or can mimic as diaphragmatic palsy and many more.[12-16] So schwannomas can involve any organ or even without involvement, they can present as a clinical illusion such that they can't be diagnosed by routine imaging like CT or MRI and only comes in picture after final histopathology report.

In conclusion, although rare, diaphragmatic schwannomas should also be included in the differential diagnosis of atypical appearing masses in the upper abdomen. Imaging like, CT scan and MRI can be equivocal and histopathology will comes as

Table 1. Showing previously reported diaphragmatic neurilemmomas

Authors	Sex	Age (years)	Side	Size (in cms)	Presentation	Treatment	Preoperative observation
Weisel et al.[3] (1956)	F	55	L	23x18	Pain, dyspnea	Thoracotomy	1 year, size increasing
Trivedi et al.[16] (1958)	F	5	L	7.5	Clubbing	Thoracotomy	None
Sarot et al.[4] (1969)	M	65	L	9x7	Asymptomatic	Thoracotomy	12 years, size increasing
McHenry et al.[5] (1988)	F	45	L	3.5	Asymptomatic	Laparotomy	None
McClenathan et al.[9] (1989)	F	46	R	10	Asymptomatic	Thoracotomy	5 years, size increasing
Koyama et al.[11] (1996)	F	38	L	5x3	Asymptomatic	Laparotomy	None
Ikegami et al.[13] (2004)	M	47	R	5	Asymptomatic	Laparotomy	13 years, size increasing
Ohba et al.[6] (2008)	F	50	R	10x8	Asymptomatic	Thoracotomy	None
Sukegawa et al.[14] (2010)	F	55	R	3.3	Pain	Thoracotomy	None
Chang et al.[15] (2012)	F	38	L	14x10	Pain, dyspnea	Thoracotomy	None
Hobbs et al.[8] (2012)	F	13	R	12x10	Pain	Laparotomy	None
Liu et al.[2] (2013)	M	47	R	3x3	Asymptomatic	Laparoscopy	19 months stable
Gungor et al.[12] (2013)	M	39	R	10	Cough, sputum	Resection	None
Present case	M	39	L	10x6	Pain	Laparotomy	None

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final diagnostic tool. Although usually benign in nature, malignant transformation had been reported, so surgical excision and follow up should be the key for proper management of such kind of cases. Despite the rare presentation of schwannomas in such forms, clinicians should have high index of suspicion with having low threshold for surgical exploration for early and proper diagnosis as well as timely management.

Informed Consent: Written informed consent was obtained from patient who participated in this case.

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