J Gastroenterol 1996; 31:268–272

Journal of Gastroenterology

Case report

Primary diaphragmatic schwannoma with a typical target appearance: Correlation of CT and MR imagings and histologic findings

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Abstract: A rare case of benign diaphragmatic schwannoma in a 38-year-old female is reported. Precontrast computed tomography (CT) showed an encapsulated well-defined round homogeneous tumor with central calcification, measuring approximately 5 cm in diameter, arising from the left diaphragm. Contrast-enhanced CT and gadolinium-enhanced T1-weighted magnetic resonance (MR) imaging showed focal enhancement in the central portion of the tumor. The tumor showed a typical target appearance of increased peripheral signal intensity and decreased central signal intensity on unenhanced T2-weighted images. Pathological examination of resected specimens of the tumor showed two zonal histological components: a hypercellular portion of spindle cells with nuclear palisading (Antoni A tissue) and a hypocellular portion of cells with cystic degeneration, together with focal calcification and hemangeomatous vascular changes (Antoni B tissue). We consider the radiological characteristics of diaphragmatic schwannoma on CT and MR imagings to represent the geographic difference between the histologic zones of the tumor.

Key words: diaphragm, Schwannoma, neurilemoma, neoplasm, CT scan, MR imaging

Introduction

Schwannoma is a benign neurogenic tumor arising from the sheath of the peripheral nerve in the soft tissues of the head and neck, extremities, mediastinum, and retroperitoneum.[1–3] Primary schwannomas of the diaphragm are rare. Precise characteristic features of the diaphragmatic tumor on radiological imagings, such as ultrasonography, computed tomography (CT), or magnetic resonance (MR) imaging have not been shown, perhaps because only a few cases have been reported.[4–8] In an attempt to characterize the CT and MR imaging findings of diaphragmatic schwannoma, we report a case of this rare tumor associated with a characteristic target appearance on radiological imagings, in which roentgenological and histopathological findings were correlated.

Case report

A 38-year-old female received a periodic examination at a local hospital and the presence of a mass in the postero-inferior left hemithorax was pointed out on a chest X-ray film (Fig. 1). The patient was referred and admitted to our University hospital in July 1994 for further workup and treatment. There was no significant past medical history; results of physical examination and routine admission laboratory studies were within normal limits. Serological tumor markers (AFP, CA19-9, CA-125, CEA, SCC, and NSE) were negative. An abdominal CT scan showed an approximately 5-cm well-defined round tumor with a relatively homogeneous area and central calcification (Fig. 2a). An enhanced CT scan showed the tumor as a double-layered zone arising from the left diaphragm, revealing a focal enhanced central-layered zone and a very low-density layered zone in the peripheral portion of the tumor (Fig. 2b). The tumor protruded into the left abdomen and its surface was smooth, without indentation. The margin of the mass was distinct. Findings on ultrasound examination were essentially analogous to those of contrast-enhanced CT. The T1-weighted MR image showed a round mass of homogeneous low signal intensity. On contrast-enhanced T1-weighted MR imaging (Fig. 3a),

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(Received for publication on May 30, 1995; accepted on Aug. 18, 1995)

however, the mass showed a high signal intensity in the central zone, with multiple degenerative areas and very low signal intensity in the peripheral zone of the tumor. The tumor had a characteristic target appearance on unenhanced T2-weighted image, with non-homogeneous decreased signal intensity in the central zone of the tumor and with markedly increased signal intensity in the peripheral zone (Fig. 3b). The radiological findings indicated that the tumor was neurogenic, such as a neurofibroma, neurofibrosarcoma, or schwannoma. To more precisely assess the tumor, ultrasound-guided biopsy was performed, but only necrotic or degenerative tissues without viable tumor cells were noted and the histological diagnosis was inconclusive. To obtain a definitive diagnosis and treat the primary diaphragmatic tumor, laparotomy and surgical excision of the tumor was performed on August 1, 1994. The mass, measuring 5.1 × 3.2 × 3.0 cm, was encapsulated, with no evidence of invasion into adjacent structures. Macroscopic examination showed the tumor cut section to be gray and yellow in color with central necrosis or degenerative changes, such as cystic degeneration, hemorrhage, and foci of calcification, and with intervening nonnecrotic tissues in peripheral portions (Fig. 4). Microscopic examination revealed the tumor to be composed of interlacing bland spindle cells with nuclear palisading Antoni A areas (Fig. 5a), and less cellular Antoni B areas associated with abundant edematous or hyalinous and collagen stroma (Fig. 5b,c). Although Antoni A and B areas were randomly mixed in the tumor, the Antoni B areas were more prominent than those of Antoni type A in the center of the tumor. Moreover, the Antoni B areas in the central zone also showed secondary changes, consisting of focal calcification and notable vascular alterations that included dilatation of the blood vessels, focal fibrinoid necrosis of the vessel walls, vascular thromboses, and hyaline thickening. However, there was no evidence of malignant change of the tumor cells. Taken together with the pathological findings, the diaphragmatic tumor was diagnosed as a typical benign neurilemoma, a so-called schwannoma. The patient had an uneventful postoperative course and is living normally.

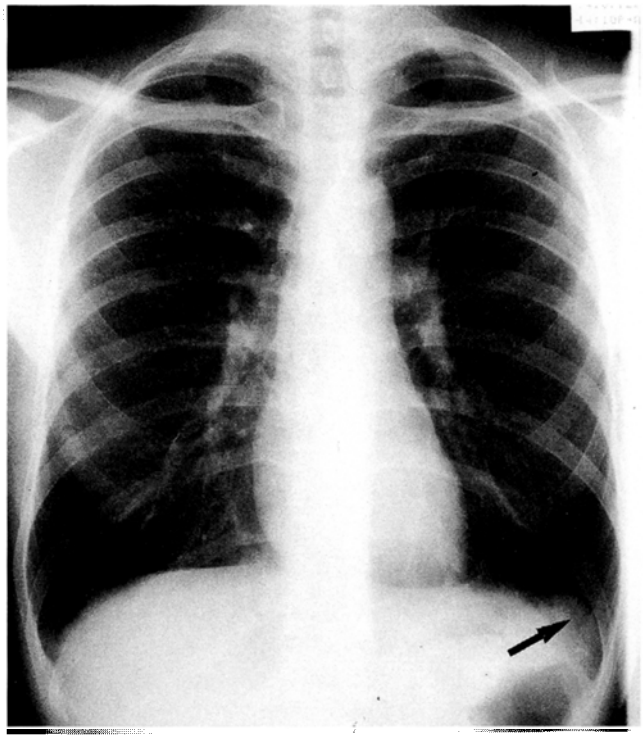

Fig. 1. Chest X-ray film showing a round tumor mass (*arrow*) adjacent to the left diaphragm

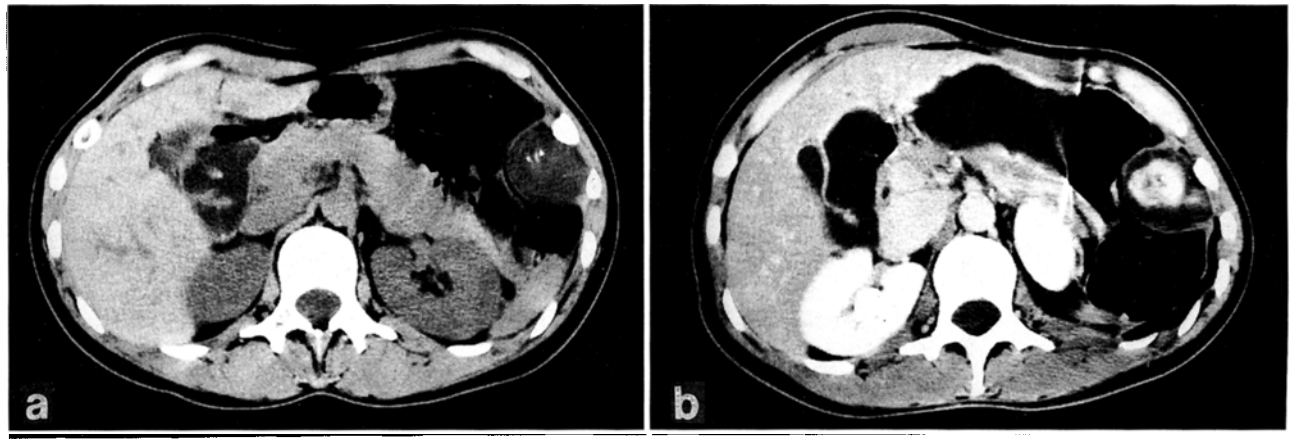

Fig. 2a,b. Abdominal computed tomography (*CT*) scan. **a** Pre-contrast CT shows a homogeneous tumor mass with central calcification arising from the left diaphragm. **b** Contrast-enhanced CT shows well-defined encapsulated tumor with focal enhancement of the central zone of the tumor

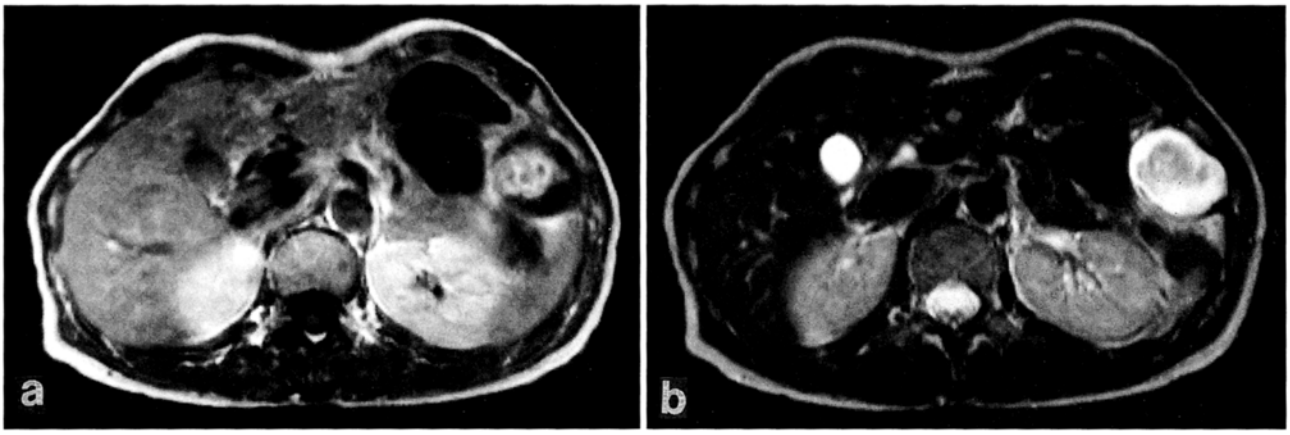

Fig. 3a,b. Abdominal magnetic resonance (*MR*) imaging. **a** Contrast-enhanced T1-weighted MR image shows a non-homogeneous round mass with high signal intensity in the central area of the tumor, and low signal intensity in the peripheral area. **b** Unenhanced T2-weighted MR image shows a typical target appearance with non-homogeneous mass and high signal intensity.

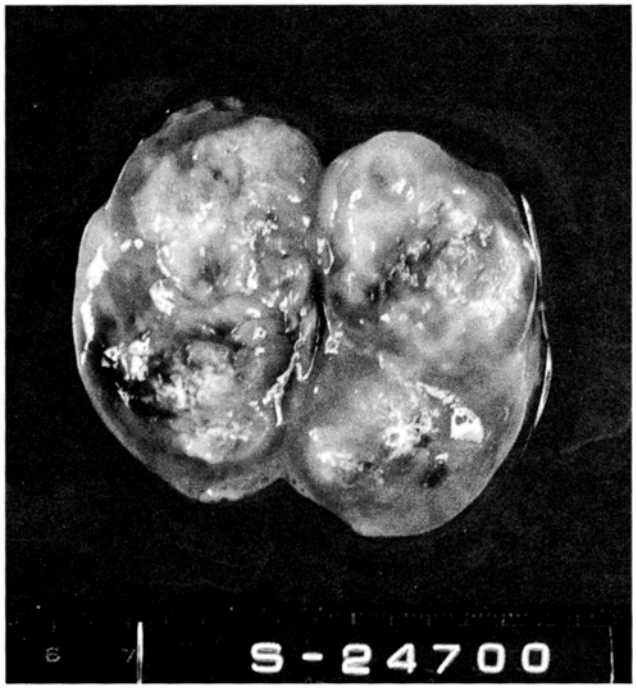

Fig. 4. Macroscopic findings of the resected diaphragmatic tumor. Cut surface of gross specimen shows necrotic or cystic degeneration and hemorrage, and intervening nonnecrotic portions

Discussion

To our knowledge, this is only the sixth case of diaphragmatic schwannoma reported in the international literature. Radiological imaging analysis showed a soft solid, calcified, and well encapsulated round mass with a typical target appearance. These findings correlated with the histological characteristics of the tumor.

Two of five previously reported cases of diaphragmatic schwannoma were analyzed by CT scan.[7,8] McHenry et al.[7] stated that CT scan disclosed a 3.5 cm uniformly enhanced mass arising from the left diaphragm. However, McClenathan and Okada[8] only described the existence of a mass originating from the right diaphragm which did not involve the liver on CT. On the other hand, Suh et al.[9] recently reported that seven of ten neurofibromas showed a typical target appearance on T2-weighted MR images, corresponding to histologically central and peripheral tissue components that differed geographically. However, they could not find a target appearance on any of the T2-weighted images examined in all four patients with schwannoma located in the neck, back or axilla. Therefore, they concluded that a mass with a target appearance on T2-weighted images most likely represented a neurofibroma, or a mass with homogeneous increased signal intensity or inhomogeneous signal intensity on T2-weighted imagings, while image without a target appearance might represent a schwannoma, neurofibrosarcoma, or atypical neurofibroma. On the contrary, our case clearly showed a typical target pattern with increased peripheral signal intensity of the tumor together with decreased central signal intensity on unenhanced T2-weighted images, despite histological confirmation of diaphragmatic schwannoma. We believe that the characteristic target appearance of the schwannoma is probably due to condensation of the T2 shorting matrix, caused by endoneural myxoid materials and dense collagen containing necrotic or degenerative tumor tissues in the central zone, as well as being due to nonnecrotic schwannoma with nuclear palisading in the peripheral zone of the tumor. Namely, the tumor in our patient showed relative zonal distinction, with the partition two zones of Antoni A and Antoni B areas on

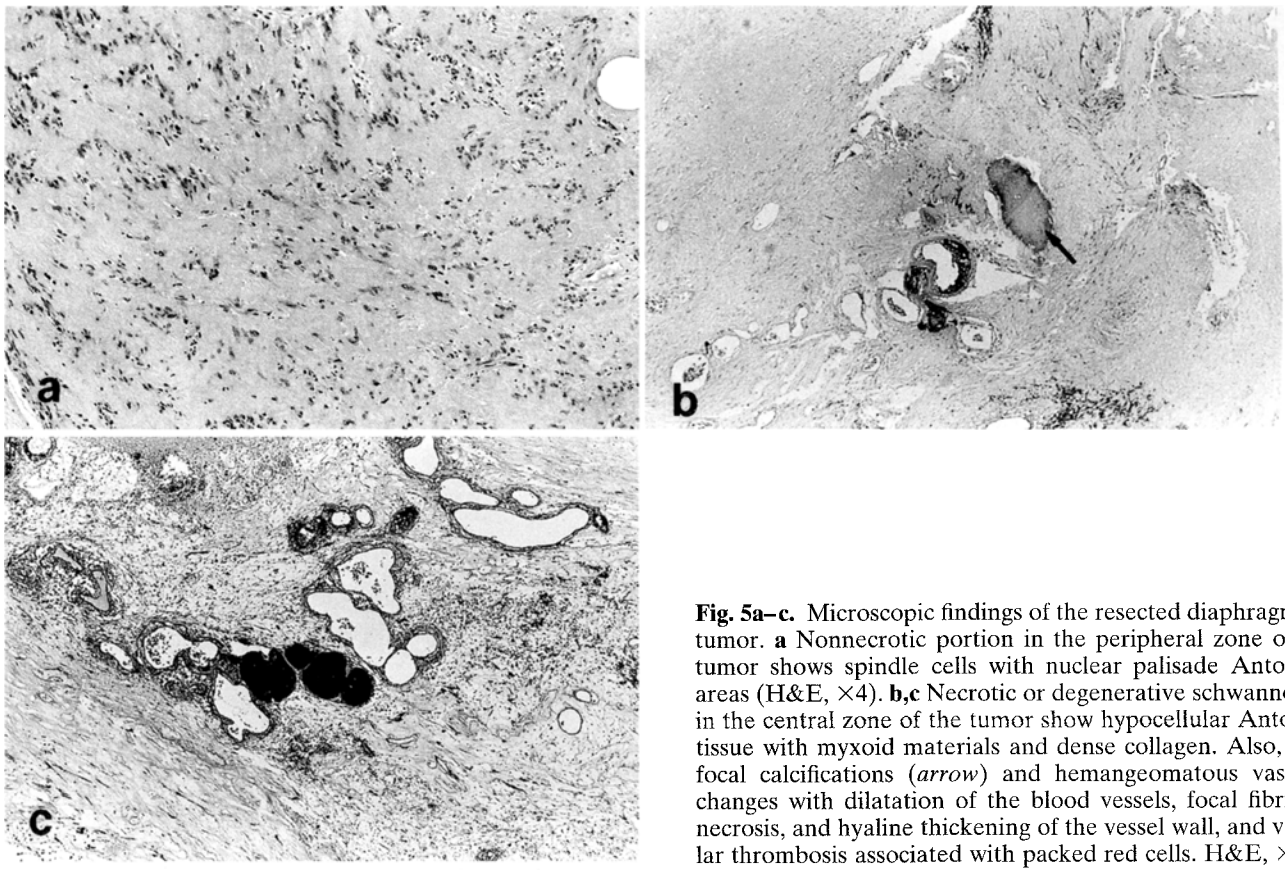

Fig. 5a–c. Microscopic findings of the resected diaphragmatic tumor. **a** Nonnecrotic portion in the peripheral zone of the tumor shows spindle cells with nuclear palisade Antoni A areas (H&E, ×4). **b,c** Necrotic or degenerative schwannomas in the central zone of the tumor show hypocellular Antoni B tissue with myxoid materials and dense collagen. Also, note focal calcifications (*arrow*) and hemangeomatous vascular changes with dilatation of the blood vessels, focal fibrinoid necrosis, and hyaline thickening of the vessel wall, and vascular thrombosis associated with packed red cells. H&E, ×3

histological evaluation, although schwannomas have been reported to usually show random distribution of the two types of tissues within the tumor.[9] Thus, it seemed likely that we had delineated the mass that appeared with a target sign typical of a benign neurofibroma, as previously described by Suh et al.,[9] on the T2-weighted MR image. Furthermore, the Antoni B area in the central zone of the tumor in this patient showed hemangeomatous vascular changes (Fig. 5b,c), suggesting increased blood flow in the lesion despite the presence of central necrosis or degenerative changes in the tumor. Thus, a focally enhanced mass within the central area of the tumor on contrast-enhanced CT and T1-weighted MR imagings (Fig. 2b and Fig. 3a) could reflect the histopathological background of the tumor. Consequently, there were correlations between the radiological features of the tumor on CT and MR images, and alternating two histological appearances of dense zonal and scanty cellular areas within the tumor.

The preoperative diagnosis of a diaphragmatic schwannoma is difficult to establish. It has been reported that needle biopsy of the tumor is an unreliable method in cases of deep-seated large schwannomas, because of the secondary histological changes of the tumor encountered in areas of degeneration in these tumors.[10,11] The results of our study agreed with the above findings and demonstrated ultrasound-guided biopsy to be inaccurate and inconclusive.

In conclusion, our study shows that contrast-enhanced CT and MR imagings provide important information for the detection of diaphragmatic schwannoma and for the preoperative evaluation of the histopathologic characteristics of this tumor.

Acknowledgments. This study was supported, in part, by a Grant-in-Aid (no. 07670560) for scientific research to S.K. from the Ministry of Education, Science, and Culture of Japan.

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