

Giant Malignant Schwannoma of the Diaphragm: CT and Ultrasound Findings

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A rare case of malignant schwannoma of the diaphragm is presented. The CT and ultrasound findings are described, correlated with the pathologic findings. During surgery, the tumor presented as a dumbbell shaped mass with central disruption of the right diaphragm.

Key Words: Diaphragm-neoplasm, ultrasound studies-neurogenic tumor-computed tomography

Primary tumors of the diaphragm are rare, particularly neurogenic tumors. Since the first report (Klassen *et al.* 1945) in 1945 of primary neurogenic tumor of the diaphragm, two cases of neurofibrosarcoma have been described (Samson and Childress 1950).

However, we have found no other reports of malignant neurogenic tumor of the diaphragm in the English literature. We present a case of malignant schwannoma that originated in the right diaphragm and discuss the imaging features in correlation with the gross pathologic and microscopic findings.

CASE REPORT

A 36-year-old woman had had pain in the right side of the lower chest and progressive cough for two weeks. In addition, she had had general weakness for two months and had lost 5 kg.

Physical examination showed diminished breathing sounds and dullness on the right lower lung field. The results of laboratory studies are normal. Chest radiograph showed a homogeneous opacification of the right lower thorax consistent with a large mass and right pleural effusion.

CT showed a large mass in the right anterior thorax, extending to the right upper abdomen. The mass was multilobulated and had multiple areas of low attenuation. Although most of the mass was located within the thorax and the heart was displaced medially, with a discernible border, a small part was within the infradiaphragmatic space and had the appearance of a tumor originating from the liver (Fig. 1A, 1B).

Combined liver-lung scintigraphy with technetium 99m showed a large, well-defined area of decreased uptake of radionuclide between lung and liver.

Sonography showed a dumbbell-shaped mass of heterogeneous echogenicity that crossed the diaphragm from the thorax to the abdomen, with disruption of the central part of diaphragm by the mass. The mass appeared to adhere to both the thoracic and the abdominal cavities (Fig. 2). Fine needle aspiration biopsies were performed on both the thoracic and abdominal parts. Examination of the specimens showed mesenchymal tissues composed of fat and some spindle cells without malignancy.

Flexible fiber-optic bronchoscopy showed a narrowed, edematous, and hyperemic bronchus in the lower lobe. The diagnosis was chronic nonspecific inflammation.

At thoracotomy, a 1560gm×10×7cm mass was removed. The right lung was displaced upward, the heart medially, and the liver downward and medially. No invasion or adhesion to these thoracoabdominal organs was seen. The diaphragmatic

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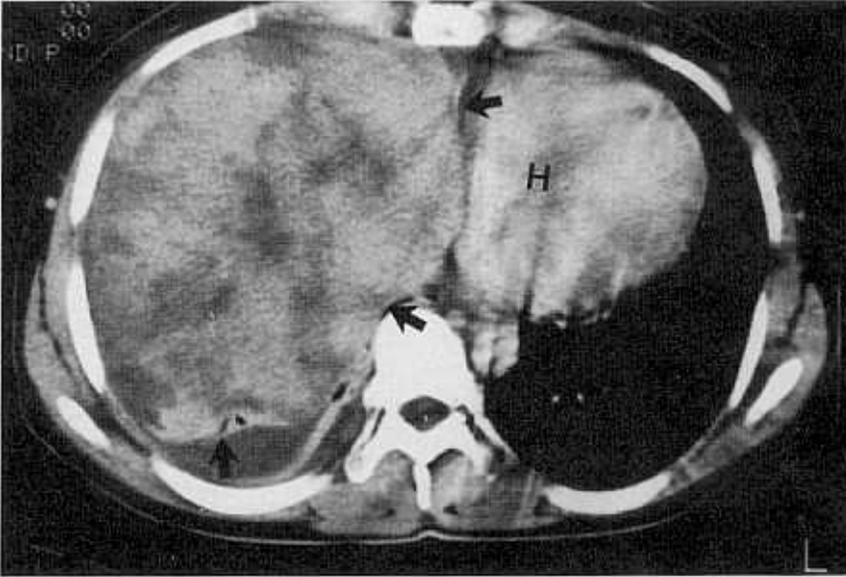


Fig. 1. A. Contrast-enhanced CT scan of thorax shows a large lobulated mass (arrows), displacing the heart (H) medially. Multiple areas of inhomogeneously low attenuation are seen within the mass which corresponded to areas of necrosis on the histopathologic examination.

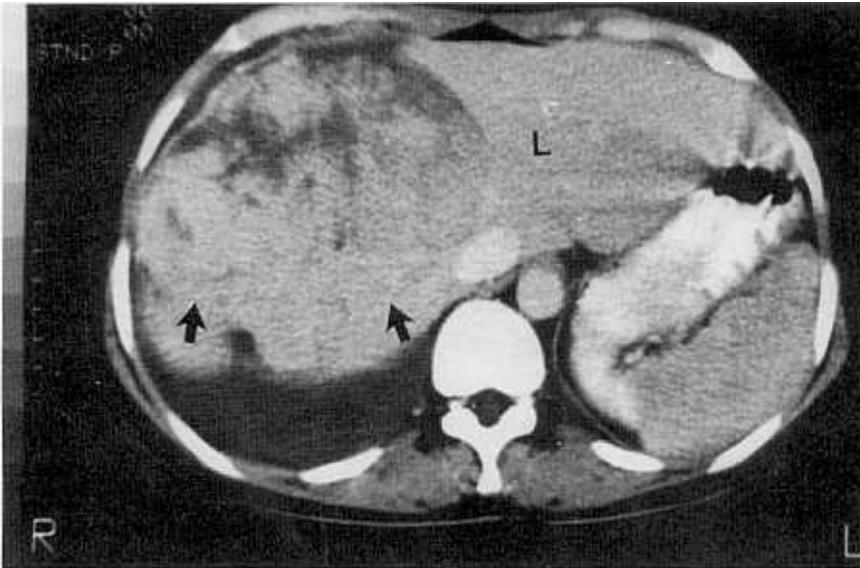


Fig. 1. B. Contrast CT scan of abdomen shows a well-defined mass (arrows), with density similar to that of thoracic mass, that appears to originate from the liver (L).

pleura adhered partially to the tumor, and the right diaphragm was disrupted. The mass was multilobulated and encapsulated, the surface was smooth

and reddish (Fig. 3). Cut sections showed a yellowish solid mass with multiple areas of necrosis and hemorrhage. Histologically the mass composed



Fig. 2. Longitudinal sonogram shows a mass with heterogeneous echogenicity, of which a part is seen in the infradiaphragmatic space (arrows), and the other in the thorax (thin arrows). The central disruption of echogenic diaphragm (white arrows) is seen (curved arrows). L : liver.

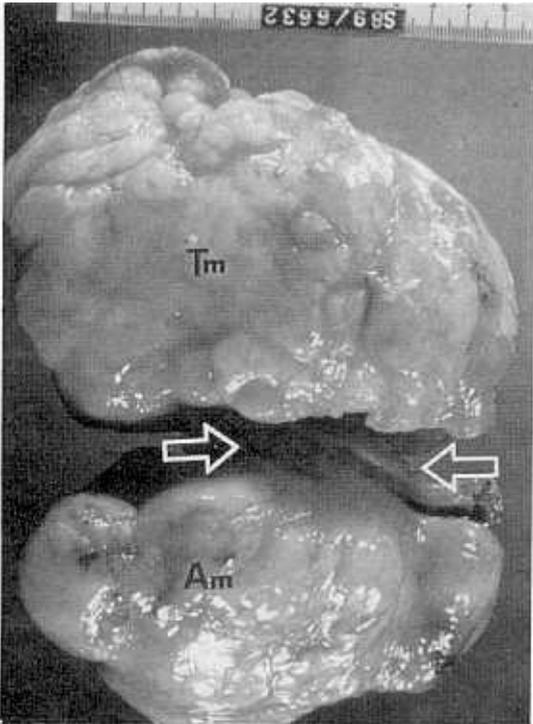


Fig. 3. Gross specimen of malignant schwannoma shows the dumbbell-shaped mass that lobulated and crossed the diaphragm (arrows) from thorax to abdomen. Tm : thoracic mass, Am : abdominal mass

mostly of long and sweeping fascicles. The nuclei were twisted, buckled, and irregular, associated with frequent mitotic figure similar to those seen in fibrosarcoma. However, immunohistochemical staining for S-100 protein was positive (Fig. 4). The diagnosis was malignant schwannoma.

DISCUSSION

All neoplasms of the diaphragm are of mesenchymal origin. Among the benign tumors, lipoma and fibroma are the most common; most of the malignant tumors are fibrosarcomas (Ackermann 1965; McHenry et al. 1988). However, primary neurogenic tumor of the diaphragm is exceedingly rare, and fewer than ten cases have been described. In 1965; Wiener and Chou (Wiener and Chou 1965) reviewed 71 cases of diaphragmatic tumors. Of these, only eight were of neurogenic origin, including two neurofibrosarcomas reported previously by Samson and Childress (Samson and Childress 1950). Although four cases of benign schwannoma of the diaphragm have been reported (McHenry et al., 1988; Cohenn et al., 1986), our case of malignant schwannoma is only the third one reported.

The CT appearance of extracranial nerve sheath tumors has been described (Cohenn et al., 1986;

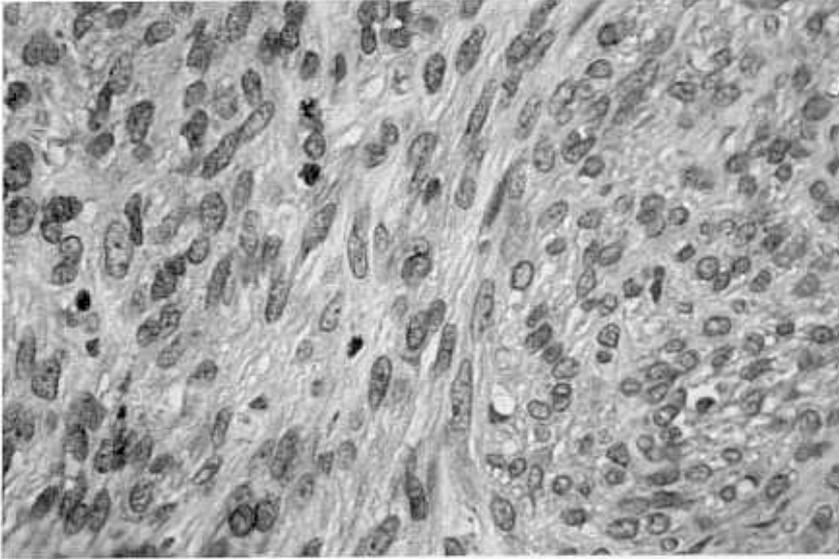


Fig. 4. Immunohistochemical staining for S-100 protein shows cytoplasmic positivity of tumor cells ($\times 400$).

Kumar *et al.*, 1983; Coleman *et al.*, 1983). Generally, benign neurogenic tumors are well-circumscribed, encapsulated, and homogeneous. Central or peripheral areas of low density are associated with the presence of Schwann cells, neural elements, adipocytes, and areas of cystic degeneration. Although areas of low density are seen in all neurogenic tumors, they are more common in schwannomas than in neurofibromas, which contain fibroblasts and nerve tissue.

On the basis of CT density, Kumar *et al.* (Kumar *et al.* 1983) classified extracranial nerve sheath tumors into two categories : those of less than 30 HU were called hypodense or lucent; those of more than 30 HU were considered dense. Masses with irregular contours that infiltrate neighboring structures or disrupt the homogeneity of the soft tissue plane, especially those showing inhomogeneously low density within solid mass, are most likely malignant (Cohenn *et al.*, 1986; Kumar *et al.*, 1983). In our case, the tumor had an irregular lobulated contour and multiple inhomogeneous areas of low density on CT, which suggest a malignant neurogenic tumor.

Sonography shows the diaphragm as well-delineated linear echogenic shadow. In our case, it clearly showed disruption of the diaphragm, which was not seen on CT. This finding suggested that the tumor

originated from or involved the diaphragm. Although the results of aspiration biopsies are not reliable enough to make a specific diagnosis, they may provide helpful information about the components of the tumor.

It is difficult to diagnose neurogenic tumor of the diaphragm on the basis of imaging features only. Accordingly the differential diagnosis should include other cystic tumors, lipoma and other malignant tumors of the diaphragm, and extradiaphragmatic tumors, especially pleural sarcoma and liver tumor (Ackermann 1942 ; McHenry *et al.*, 1988 ; Wiener and Chou 1965).

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