ورم الغمدة العصبية الخبيث للمنصف الخلفي Malignant Schwannoma of posterior mediastinum A Case report and review of literature

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Malignant Schwannoma of posterior mediastinum A Case report and review of literature

SUMMARY:

In this report rare case of malignant schwannoma of posterior mediastinum was reported in 55 years old man, who's his Magnetic Resonance Images and computerized axial tomography scan of the chest revealed two masses in posterior medistinum measured 12.4 X13.8cm, 11X7cm located on both side of aorta, with smooth outline, encasing the descending aorta posterior to the heart.

Open Biopsy of the lesions revealed malignant schwannoma.

الخلاصة

في هذا التقرير يتم تسجيل حال نادرة وهي ورم الغمدة العصبية الخبيث للمنصف الخلفي في مريض يبلغ من العمر 55 عاماً وحيث أظهر فحص الرنين المغناطيسي و المفراس الحلزوني للصدر وجود ورمتين في المنصف الخلفي ذي أبعاد 11X7 و 12.4 X13.8 وعللا جانبي الابهر النازل وخلف القلب. الخزعة الجراحية المأخوذة من الآفة أظهرت ورم الغمدة العصبية الخبيث

key word: Schwannomas, neurinomas, neurilemomas, neurolemomas, diagnosis MRI feature, Malignante changes.

Introduction:

Schwannomas are also called neurinomas, neurilemomas, neurolemomas.^{1, 2} They arise from Schwann cell of nerve root most frequently the 8th cranial nerve (acoustic schwannoma), the fifth cranial nerve is the second most frequent site.¹

The tumour histologically characterized by the proliferation of Schwann cells that arise from the neural sheath of autonomic, cranial or peripheral nerves ². The tumor usually shows spindle-cell proliferation in a concentric or fascicular pattern. ³ Immunohistochemically the tumor cells are diffusely positive for S-100 protein, and they have intracytoplasmic periodic acid Schiff (PAS)-positive crystalloids. ³

The tumors are usually begnin but in rare exception malignant transformation may occur. ¹they are very rarely occur out side the head and neck region.

In this report malignant schwannoma of posterior mediastinum was reported, through light on it's presentation and the way of diagnosis with review of literature

Case report

On 14 of April 2002, 55 years old man referred for echo clinic for assessment of left ventricular function, because of shortness of breath on exertion, prolong chest pain and cardiomegally on chest x-ray. On review of his history, he had progressive shortness of breath for last 3 month, this was associated with chest pain which was prolong, lasting hour or more mainly in the retrosternal area with radiation to right side of the chest and back .No orthopnea, paroxysmal nocturnal dyspnea, or legs swelling. A review of other systems were not revealed any significant symptoms

On examination: He was well build, not dyspneic, and not cyanotic. His Jugular venous pressure was normal. Pulse rate was 80\minute and regular. His blood pressure was 140\ 80 mmhg. Chest examination revealed dullness and diminished breath sound at mid and lower right zone. On cardiac auscultation, there was normal first and second heart sound with no added sound. His chest X Ray show big homogenous mass occupying the right lower and mid zone obliterating the right cardiac silhouette (Figure 1) .on lateral veiw the mass occupying the posterior medistinum. Echocardiography showed normal cardiac size and normal LV Function.

Magnetic Resonance Images (MRI) of the chest showed big mass with heterogeneous intensity in T2 and hypointesity in T1, located in posterior medistinum (Figure2)

Computerized axial tomography scan of the chest (Figure 3, 4,) reveal two masses in posterior medistinum measured 12.4 X13.8cm, 11X7cm located on both sides of aorta, with smooth outline, encasing the descending aorta post to the heart these masses were not enhanced by contrast media. The radiologist suggested begnin nature of the mass according to the above features. Both CT scan and MRI angiography showed that there was no evidence of aortic aneurysm. (Figure 4,5)

On 22nd of April 2002, Thoracotomy was done revealed huge post mediastinal tumor posterior to the heart and encasing the descending aorta. It was unresctable. Biopsy was taken and submitted to histopathological examination, which revealed malignant Shwanoma. patient received five session of radiotherapy ,despite he felt well after that, but the radiological assessment of the masses didn't show any improvement in the size of the tumor.

Discussion:

The peripheral nerve tumors are of two types, neurofibromas and schwannomas. Neurofibromas are the commonest form. It may arise from sensory nerve twigs produce the distinctive subcutaneous nodules; in peripheral nerve trunks the tumor appears as a fusiform enlargement or plexiform neuroma.⁴

Schwannomas are rare and arise from Schwann cell of nerve root most frequently the 8th cranial nerve (acoustic schwannoma), the fifth cranial nerve is the second most frequent site.¹

There are reports of rare cases of malignant schwannoma of the gastrointestinal tract ^{2,3}, ovary, peritoneum or urinary bladder. ^{6,8}

There are only few reports of rare cases of malignant schwannoma which involve the posterior mediastinum^{5,7}

In the vast majority of cases, schwannoma of the posterior mediastinum arises from one of the intercostal nerves and most often is manifested by an asymptomatic solitary mass on a radiograph. 5

In this reported two large masses of posterior mediastinum were detected measuring 12.4 X13.8cm, 11X7cm, this was consistent with Hayasaka-K et al finding that a malignant lesion size usually above 10 cm ,and benign schwannomas usually have: tumor diameter of 5.5 + 3.1 cm.⁶

Chest x ray was good tool for localizing the opacity to posterior mediastinum but CT scan and MRI were more informative⁸ in deciding the nature of opacity and its exact anatomical origin and it's relation to adjacent structures. In this case both image exclude the possibility of aortic aneurysm or esophageal origin of the mass since these two are common causes of posterior mediastinum masses ^{4,8}

An interesting observations are these images showed that the lesions had smooth margin, finding that consistent with begnin nature of the lesions radiologically ⁶. Hayasaka-K administered that the tumor margin was regular in all of the benign cases and irregular in the malignant one, ⁶ But in this case, it was malignant on histopathological examination despite smooth margin on MRI and CT scan. This suggests that this radiological feature is not constant one.

These masses on MRI had heterogeneous intensity on T2 and hypointensity on T1. Hayasaka-K administered that the different signal intensity may be due cystic degeneration⁶, however there is no specific signal characteristics of these tumors⁶

Conclusion:

Malignant schwannoma of posterior mediastinum is quiet rare, CT scan MRI good tools for diagnosis but the begnin or malignant nature of the tumor only can confirm by histopathological examination

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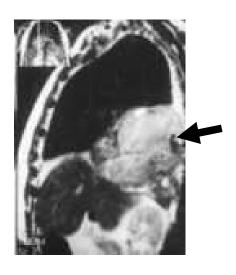


Figure2 posterior mediastinal mass

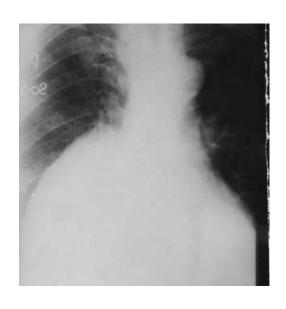


Figure chest X ray give the impression of cardiomegally



Figure 3 CT scan chest the masses are not aneurysm





Figurer 5 MRI show the two masses encase the aorta