RARE PRESENTATION OF A CASE OF GANGLIONEUROMA (A Case Report)

Lt Col PK GUPTA^{*}, Lt Col VK BATISH⁺,
Lt Col GU DESHPANDE[#], Col SK KHANNA^{**},
Col AMARJIT SINGH⁺⁺

MJAFI 1997; 53 : 82-84

KEYWORDS :

Introduction

Ganglioneuroma is an uncommon benign tumor commonly seen between the age of 10-30 years. It is invariably encountered as a mediastinal mass on chest radiograph. Patients are usually asymptomatic and normotensive. The tumor contains mature ganglion cells. It rarely turns malignant even after decades of benign existence [1-3]. Imaging modalities give a definite clue to the existence of a neurogenic mass in the posterior mediastinum but it is finally diagnosed on histopathology examination.

Case Report

A 5-year-old female patient was brought to the hospital for chest deformity and thin built otherwise asymptomatic. On examination the child was normotensive with normal physical, emotional and mental development. The child had minimal pectus excavatum, cafe au lait spot on the right anterior lower chest wall. Biochemical parameters were within normal limits. Chest radiograph showed a triangular mass in the posterior mediastinum having the base towards the mediastinum. The right thoracic cage showed ribbon shaped 5th to 8th ribs with thinning and erosion (Fig 1). Anterior notching of the thoracic vertebral bodies D5 to D8 were seen on lateral radiograph of the thoracic spine. Ultrasound of the abdomen was done to rule out any other abnormality. It revealed a right suprarenal mass with areas of necrosis within. No calcification was detected. Computerised axial tomography (CT) done showed an enhancing mass in the thorax mainly on the right side with extradural extension into the spinal canal displacing the cord to the left. Mass was seen to extend along the posterior aspect of the lower thoracic ribs. There was a diffuse angioma under the cafe au lait spot on the right lower chest and in the erector spinae muscle posteriorly on right side. There was a minimally enhancing abdominal mass with both crus of diaphragm in normal location (Fig 2). The right kidney was free from the mass but displaced downwards. Pre-operative magnetic resonance imaging (MRI) was done to evaluate the complete intra spinal extension on the mass. MRI showed an extradural mass with compression of spinal cord (Fig 3). Based on these findings a diagnosis of

neurofibroma was made.

Histopathological examination of the specimen obtained following spinal surgical excision revealed tumor tissue comprising of longitudinally and transversely oriented Schwann cells criss crossing each other in an irregular fashion. Scattered throughout these cells were mature ganglion cells which were seen lying singly and in clusters. Histopathological examination thus confirmed the diagnosis of the ganglioneuroma (Fig 4).

Discussion

Ganglioneuroma is a rare tumor in childhood especially at the age of 5 years. Usually it is located in the

Fig. 1 : Chest PA view shows a triangular soft tissue mass on the right side. The right 5th to 8th ribs are ribbon shaped with thinning of the thoracic vertebral pedicles at the same level.

^{*}Classified Specialist Radiology and Nuclear Medicine, ⁺Classified Specialist in Surgery and Neurosurgery, ^{**}Senior Advisor in Radiodiagnosis, Command Hospital (SC), Pune 411040; [#]Reader in Pathology, ⁺⁺Professor and Head of Radiodiagnosis, Armed Forces Medical College, Pune 411040

Fig. 2 : Abdominal CT scan showing non enhancing soft tissue mass in the right supra renal region displacing the IVC. Anterior abdominal wall on right is enhancing at the site of cafe au lait spot.

Fig. 3 : T1 weighted MRI sagittal section of the spine showing the compression of the cord; anterior notching of the thoracic vertebrae and the mediastinal soft tissue mass.

posterior mediastinum though abdominal masses separately are also described, but presence at both sites in the same patient is extremely rare as seen in this patient.

Ganglioneuroma arise from the ganglion cells and contain mature ganglion cells in cluster and nests against the background of the longitudinal and transversely oriented schwann cells [1]. In the study by Heimbürger et al the posterior mediastinal tumors were 24 per cent of all the mediastinal tumors of which 82 per cent were of neurogenic origin and of

Fig. 4 : Schwann cells arranged in fascicles with ganglion cells in the center.

these 80 per cent were from sympathetic chain [3]. Ganglioneuroma is an uncommon tumor and was seen in only 11 patients out of 186 mediastinal masses in the series by Rubush et al [4], and 2 out of 42 cases in the series by Heimbürger [3]. The ganglioneuroma cases mostly present after the age of 10 years [4], only 9 patients of 88 cases were between 5-9 years [1]. Patients are usually asymptomatic, normotensive and may be rarely associated with neurofibromatosis, Von Recklinghausen's disease, myasthenia gravis, virilisation, and adrenal involvement. Occasional diarrhoea and hypotension may be present [1,4]. The lesions are most commonly located in the posterior mediastinum or retroperitoneal region. Presence at both the locations in the same patient is not documented in the literature reviewed. Chest radiograph shows a paravertebral triangular mass with base ribbon at the mediastinum on postero-anterior view while lateral chest radiograph shows posterior location of the mass with superior and inferior margins remaining indistinct. Ribbon shaped ribs and vertebral pedicle erosion have also been described. The latter indicates intraspinal extension.

Sonography evaluation of these soft tissue masses reveal hypoechoic areas which indicate necrosis [1]. In about one third cases calcification is seen [1] but was not present in this case either on ultrasound examination nor on CT evaluation. The hour glass extension also known as dumb bell tumor extending into the spinal canal through neural foramina is rare [4], was

well seen in this patient. These lesions are well appreciated on CT while MRI is useful in post operated cases including the recurrence, post radiation therapy evaluation and preoperatively where intraspinal extent is to be evaluated [2]. These tumors rarely turn malignant for decades [1]. Ganglioneuromas may also be a sequel of the maturing up of the neuroblastoma in adulthood survivors [6]. One case of ganglioneuroma arising from adrenal in HIV positive patient transformed into malignancy has been documented [7].

REFERENCES

1. Enzinger MF, Weiss WS. Primitive Neuroectodermal Tumors and Related Lesions. In: Enzinger MF, Weiss WS, editors. Soft Tissue Tumors. 3rd ed. USA: Mosby, 1995; 928-64.
2. H Dirk Sostman, Webb WR. MRI in Thoracic Diseases. In: Potchen EJ, Graninger RG, Greene R, editors. Pulmonary Radiology by Members of Fleischner Society. Pennsylvania WB Saunders, 1993; 361-78.
3. Heimbürger IL, Battersby JS. Primary Mediastinal tumors. J Thoracic Cardiovasc Surg, 1965; 50: 92-103.
4. Rubush JL, Gardner IR, Boyd WC, Ehrenhaft JL. Mediastinal Tumors Review of 186 Cases. J Thorac Cardiovasc Surg, 1973; 65: 216-22.
5. Davis RD Jr, Oldham HN Jr, Sabiston DC Jr. The Mediastinum. In: Sabiston DC Jr, Spencer FC, editors. Surgery of Chest Vol 1. 6th ed, Pennsylvania: WB Saunders, 1995; 76-611.
6. Grossman A. Adrenal tumors. In: Peckham M, Pinedo H, Veronesi U, editors. Oxford Textbook of Oncology Vol 2. London: Oxford University Press, 1995; 137-2145.
7. Chandra SP, Shibata D, Radin R et al. Malignant Peripheral Nerve Sheath Tumor Arising in an Adrenal Ganglioneuroma in an Adult Male Homosexual. Cancer 1986; 57: 2022.