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CASE REPORT
ΕΝΔΙΑΦΕΡΟΥΣΑ ΠΕΡΙΠΤΩΣΗ

Radiotherapy for paravertebral ganglioneuroma

Ganglioneuroma is a tumor of sympathetic ganglion origin, located in the paravertebral area, consisting of mature ganglion cells, and it is homogeneously encapsulated. It is generally seen in children and young adults. Its treatment is complete surgical resection. Due to the benign nature of the tumor, adjuvant systemic chemotherapy and local radiotherapy have a limited role in its treatment. The case is reported here of a 46-year-old male in whom a mass located in the thoracic region was detected on computed tomography (CT) performed for back pain. Ganglioneuroma was diagnosed, and the patient underwent subtotal resection of the tumor, followed by 30 Gy (3 Gy fraction/day) radiotherapy, because of incomplete resection. There has been no progression in the 4-year follow-up of the patient. The clinical features, site of the mass, treatment approaches, and the importance and effectiveness of radiotherapy in a patient with ganglioneuroma located in the thoracic region are discussed.

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Ακτινοθεραπεία σε παρασπονδυλικό γαγγλιονεύρωμα

Περίληψη στο τέλος του άρθρου

Key words

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Radiotherapy
Thoracic vertebra

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Ganglioneuroma is a rare, benign tumor consisting of sympathetic ganglion cells.[1] It may occur spontaneously, or as the maturation of neuroblastoma, or as a metastasis after chemotherapy or radiotherapy. It develops along the sympathetic chain, extending from the base of the skull, neck, posterior mediastinum, retroperitoneum, and to the adrenal gland.[2–4] It is most commonly observed in the posterior mediastinum and the retroperitoneum. It is usually diagnosed in childhood.[1,2] Since the tumor is not metabolically active, it is usually asymptomatic and is rarely detected before it reaches a large size. Symptoms are usually due to pressure of the tumor on the surrounding tissue, often the nerves. In addition to neural compression, dorsal spinal scoliosis, and symptoms due to increased catecholamine secretion may be seen, rarely.[3] The diagnosis is made by magnetic resonance imaging (MRI) or computed tomography (CT).[4]

Treatment is excision of the total mass, whenever possible. Adjuvant systemic chemotherapy and local radiotherapy are of limited effectiveness due to the benign biological nature of the tumor. The prognosis is good for those who have undergone total tumor excision, but local recurrence may occur after surgical resection, especially subtotal resection, and routine follow-up is recommended even after complete excision.[1] The clinical behavior, diagnosis and the treatment of paravertebral ganglioneuroblastoma are discussed.

CASE PRESENTATION

A 46-year-old male presented to the chest diseases outpatient clinic with back, neck and chest pain of about a month's duration. He had a smoking history, but nothing of significance in his medical and family history. On physical examination there was no finding other than tenderness in the costovertebral region. On chest X-ray, a well-circumscribed, round lesion was observed at the base of the left first rib. Routine laboratory tests were within normal limits. Thoracic CT revealed a mass of 6×6 mm in size in the lung noddle area in the anterior segment of the right lung upper lobe, and a lesion adjacent to the T6–T11 vertebrae in the right hemithorax (fig. 1A). Spinal MRI showed a 130×105×61 mm mass, starting from the inferior level of the T6 vertebra, filling the right paravertebral area, and compressing the spinal cord (fig. 1B).

The patient underwent surgery based in the provisional biopsy diagnosis of a benign mesenchymal. It was decided during the operation to perform thoracotomy, as it was thought that the tumor was neurogenic. Debulking surgery was performed on the mass,

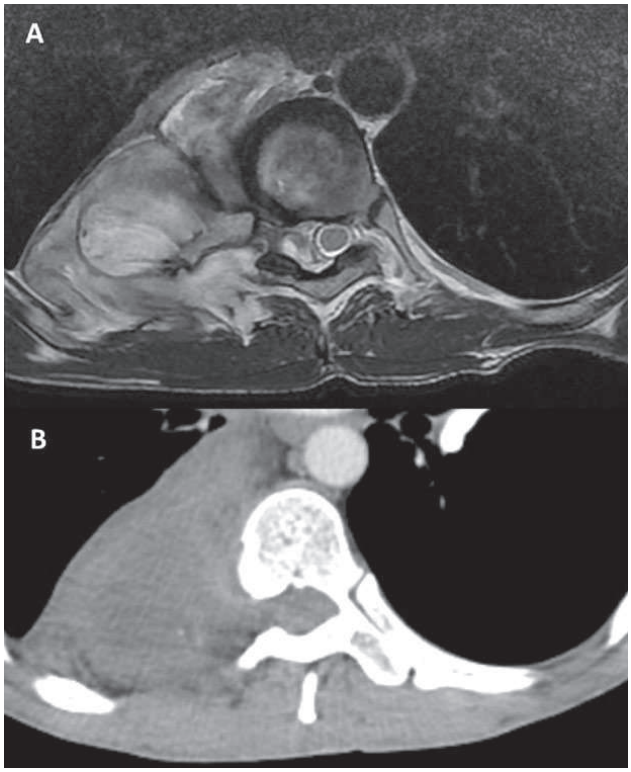

Figure 1. Ganglioneuroma in a 46-year-old male. Preoperative computed tomography (CT) (A) and axial contrast-enhanced T2W magnetic resonance imaging (MRI) (B) show a mass in the right paravertebral area extending into the neural foramen and spinal canal.

and wedge resection was performed for the nodule in the right lung. No complications were observed in the postoperative period. The histopathological examination showed a paraspinal ganglioneuroma (NSE ganglion cells +, S100 and stroma surrounding ganglion cells +), and emphysematous changes in the lung mass.

As the lesion was benign, it was decided to monitor the patient. At 3-month follow-up, spinal MRI showed an irregularly enhancing mass sized 120×75×30 mm, extending to the posterior cords in the right paravertebral area at the level of the T7–T11 vertebrae (figures 2A, 2B), and a consultation was held between the neurosurgery and thoracic surgery departments. As the tumor was considered to be inoperable, and the patient was in intense pain and developed scoliosis thirty gray (3 Gy fraction/day, 10 fractions) radiotherapy was applied. The patient has been followed for 4 years and no progression is observed (figures 2C, 2D).

DISCUSSION

Neurogenic tumors account for 20% of all mediastinal tumors in adults and concern the most common cause of posterior mediastinal masses.[2] Neurogenic tumors are grouped according to their origin from peripheral nerves, sympathetic ganglia or parasympathetic ganglia. The International Neuroblastoma Pathology Classification classifies them in four categories: neuroblastoma, ganglioneuroblastoma intermediate, ganglioneuroblastoma nodular and

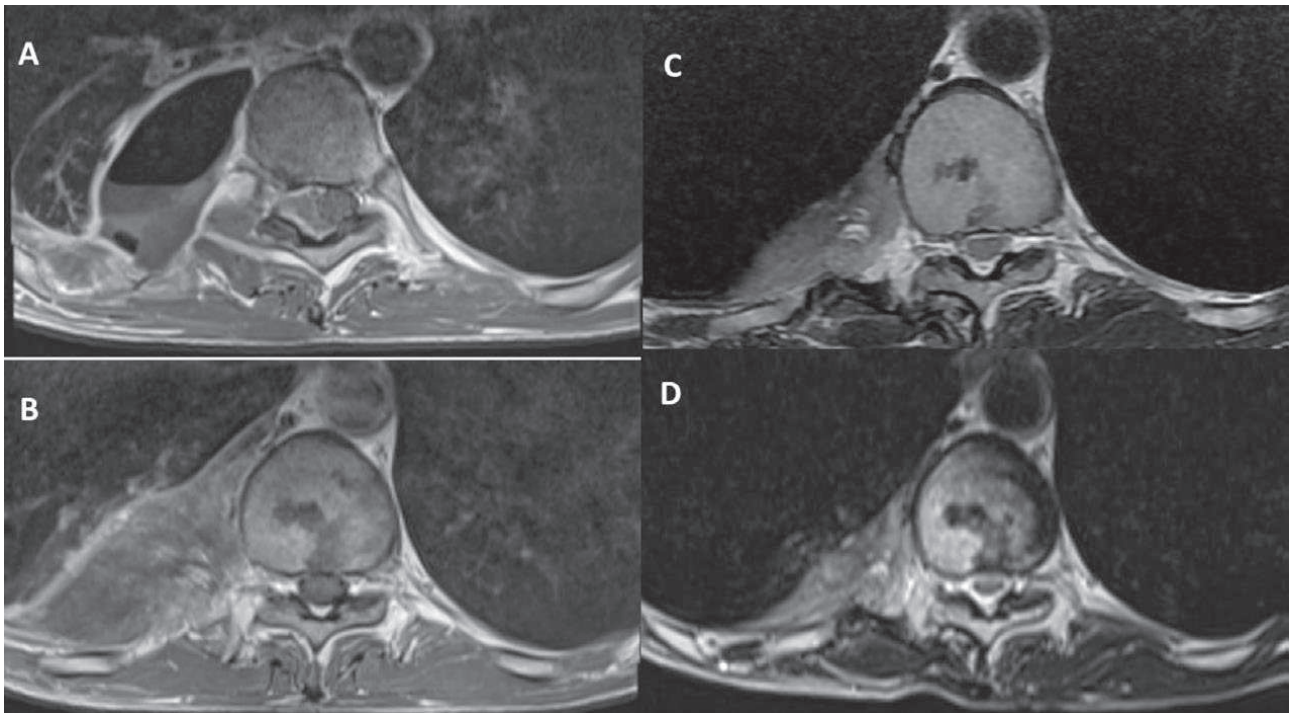

Figure 2. Ganglioneuroma in a 46-year-old male. Axial contrast-enhanced T1W magnetic resonance imaging (MRI) shows the remaining tumor after subtotal surgery (A) and an increase in size of the residuol tumor 3 months after surgery (B) Axial T2W MRI shows the lesion on the first year (C) and the 4th year after radiotherapy (D).

ganglioneuroma.[4] Ganglioneuroma, which is a sympathetic ganglion tumor, is an encapsulated, homogeneous benign tumor consisting of single or clustered mature ganglion cells.[1] It originates mainly from sympathetic ganglia in the posterior mediastinum and may spread intraspinally.[6–8] Because of its slow growth, it is usually diagnosed in adolescence. The symptoms are usually caused by the compression of the tumor on the surrounding tissue. In a series reported by Reed et al, there were only two patients aged between 40 and 50 years among 38 patients with ganglioneuroma, and no patient over the age of 50 years.[9] In the series of Ribet and Cardot, similarly, only two of 35 patients with ganglioneuroma were aged between 45 and 54 years.[10] In our case, the patient was aged 46 years old at diagnosis, by which time the mass had reached large dimensions that caused back pain, which disappeared after surgery. No biochemical or metabolic disturbances were observed.

Nowadays, the incidence of diagnosis of ganglioneuroma has increased, due to the ease of access to imaging studies. Preoperative diagnosis may be difficult because such masses are radiologically similar to other tumors. Chest X-ray, CT and MRI are the imaging studies that lead to clear visualization of the position and extent of the tumor. It is often seen on chest X-ray as a lesion located on the anterolateral edge of the vertebral canal and separated from the surrounding tissues by sharp boundaries. CT is valuable in detecting the size, location and the bony changes of the tumor, but MRI is preferred in the case of extension, because it shows the relationship between the tumor and the spinal cord.[4,6–8] In our case, all three imaging techniques were used to ensure more accurate visualization.

The histopathological diagnosis is usually made by needle aspiration or open biopsy. The primary treatment is removal of the tumor, complete whenever possible. The definitive diagnosis is made by cytological and immunohistochemical studies after the surgical excision.[1,2] In our case, a diagnostic biopsy was performed to determine the localization and the nature of the mass. No intraspinal dissemination was observed and subtotal surgical resection was performed.

In conclusion, when the literature was reviewed, this case was found to be important, because ganglioneuroma is rare over the age of 40 years, and the treatment approach is different from that in younger patients. In this patient, radiotherapy was applied in the hope that it might be effective, as recurrence developed after subtotal resection, and the patient could not be operated on a second time. Although the initial surgical treatment was incomplete, a long-term disease-free interval was observed as a result of radiotherapy.

ΠΕΡΙΛΗΨΗ

Ακτινοθεραπεία σε παρασπονδυλικό γαγγλιονεύρωμα

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Το γαγγλιονεύρωμα είναι όγκος του συμπαθητικού αποτελούμενος από ώριμα γαγγλιοκύτταρα, τα οποία εντοπίζονται στην παρασπονδυλική περιοχή και περικλείονται ομοιογενώς σε κάψα. Εμφανίζεται σε παιδιά και νεαρούς ενήλικες. Η θεραπεία του είναι η πλήρης χειρουργική εκτομή. Η επικουρική συστηματική χημειοθεραπεία και η τοπική ακτινοθεραπεία διαδραματίζουν περιορισμένο ρόλο λόγω της καλοήθους φύσης του όγκου. Περιγράφεται η περίπτωση ενός άνδρα, ηλικίας 46 ετών, που παρουσίασε άλγος στην πλάτη και στην αξονική τομογραφία εντοπίστηκε μια παρασπονδυλική μάζα στη θωρακική περιοχή. Τέθηκε η διάγνωση γαγγλιονευρώματος και ο ασθενής υποβλήθηκε σε υφολική εκτομή του όγκου. Χορηγήθηκε επίσης ακτινοθεραπεία 30 Gy (3 Gy/ημέρα). Δεν υπήρξε υποτροπή της νόσου τα τελευταία 4 έτη. Στο παρόν άρθρο αναφέρονται τα κλινικά χαρακτηριστικά, οι θεραπευτικές προσεγγίσεις της θεραπείας, η θέση του όγκου, καθώς και η αποτελεσματικότητα και η σημασία της ακτινοθεραπείας στο γαγγλιονεύρωμα της θωρακικής περιοχής.

Λέξεις ευρετηρίου: Ακτινοθεραπεία, Γαγγλιονεύρωμα, Θωρακικοί σπόνδυλοι

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