Paraspinal ganglioneuroma

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Neurosurgery Department, University General Hospital of Alicante, Foundation for the Promotion of Health and Biomedical Research in the Valencian Region (FISABIO), Alicante, Spain Spinal ganglioneuroma represent less than 10% of all ganglioneuromas, usually with the involvement of the paraspinal region [1].

Most commonly they arise in the paravertebral sympathetic chains of the posterior mediastinum (41.5%) or retroperitoneum (37.5%). Less common sites include the adrenal gland (21%) and neck (8%) [2].

Classification

In 1941, Eden classified dumbbell-shaped tumors into four categories according to the anatomical relationship, that is, the spinal cord and vertebrae: intradural and extradural; intradural, extradural, and paravertebral; extradural and paravertebral; and foraminal and paravertebral [3].

It is extradural and paravertebral according to the Eden categorization. The most common dumbbell-shaped tumor is the Schwann cell tumor, whereas ganglioneuroma is relatively rare [4].

Paravertebral ganglioneuromas frequently extend through the neural foramina to involve the epidural space of the spinal canal. Intradural extramedullary ganglioneuromas have been reported but are extremely rare [5].

Clinical features

Ganglioneuromas can remain asymptomatic for a long period and can be diagnosed incidentally.

Clinically, radicular pain and sensory-motor disorders are the main manifestations. Its diagnosis depends on pathological examination. [6].

Diagnosis

At MR imaging, GN has low signal intensity on T1-weighted images and high signal intensity on T2 weighted images [7]. Distinct radiological features are lacking. Therefore, diagnosis can be challenging and is more precisely achieved by histological examination after resection. An imaging-guided core needle biopsy can be a reasonable approach to enable a reliable diagnosis before major surgery.

Radiographic features

Plain radiograph

may show a posterior mediastinal mass, sometimes causing rib spreading and foraminal erosion a retroperitoneal mass may be identified scoliosis may be present.

CT

homogeneous low attenuation mass

calcifications are seen in 42-60%

calcification is typically fine and speckled but may be coarse

slight to moderate enhancement which may be heterogeneous or homogeneous

MRI

Ganglioneuromas are well circumscribed masses. They may appear encapsulated, although a true capsule is infrequent 3. Signal characteristics include:

T1: low signal intensity may have a whorled appearance

T2: heterogenous high signal intensity

T1 C+ (Gd): enhancement varies from mild to marked

Differential diagnosis

spinal neuroblastoma and ganglioneuroblastoma often cannot be differentiated from ganglioneuroma based on imaging characteristics metastases usually indicate neuroblastoma or ganglioneuroblastoma compared with ganglioneuromas, tumour calcification in neuroblastomas is more often amorphous and of a rough pattern spinal schwannoma and neurofibroma centred on neural foramen primarily seen in middle-aged adults

Treatment

Surgical excision is the treatment of choice in the management of symptomatic ganglioneuromas with very good prognosis [8)].

Outcome

They may grow to a large size. Despite the size and the common involvement of both intra- and extraspinal compartments, the prognosis is usually excellent after complete excision using microsurgical techniques [9)]

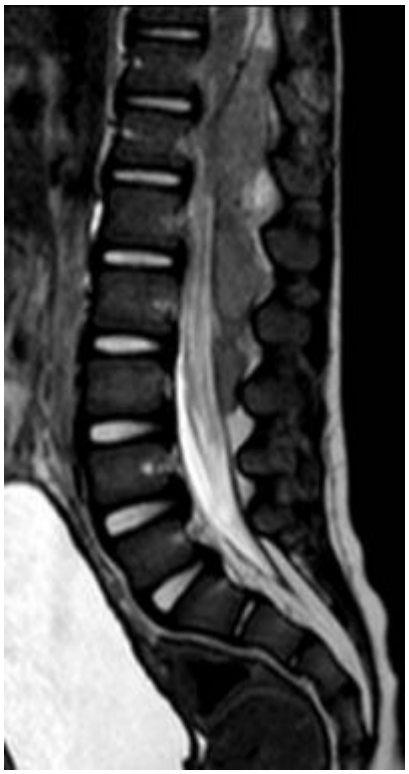
see Conus medullaris ganglioneuroma.

Case report

A 2-year-old girl with recurrent urinary infections, persistent constipation from 4 months of age with rectal prolapse and congenital metatarsal foot supine deviance of the feet inwards.

In the context of a study of the chronic condition described, the patient was requested to rule out spina bifida.

Tumor of soft tissues of extradural location Located between D11 and L3, which extends out of the canal through conjunctival foramen L1-L2, L2-L3 and L3-L4 both to the right paravertebral musculature and to the medial space to the homolateral kidney. Pathological left paraaortic retroperitoneal lymphadenopathy.



Case series

The clinical data of 6 patients underwent a surgery for ganglioneuroma in spine from January 2008 to January 2015 were retrospectively analyzed. There were 4 males and 2 females, aged from 2 to 63 years old with an average of 34.6 years. The courses of disease were from 3 days to 17 years. Five patients complicated with superficial hypesthesia in correlative level of tumor, and the muscle strength under tumor plane had decreased at different levels, with the strength of grade II-IV. Two cases complicated with hypermyotonia and positive bilateral Hoffmann's and Babinski sign. Five cases were sporadic lesion in correlative spinal canal and one case complicated with the giant occupying lesion in thoracic cavity.

Six operations had been performed including 5 en bloc and 1 subtotal resection. Postoperative pathological results showed tumor cells scattered or fasciculated inserted into Schwann cells in the stroma. In 2 patients complicated with radiculalgia before operation, 1 case was relieved and 1 was invariant after operation. All 4 patients with preoperative dyscinesia in the limbs obtained improvement after operation. All the patients were followed up from 0.3 to 6.8 years with an average of 2.5 years. At the final follow-up, according to ASIA grade, 5 cases were good and 1 case was invariant. During the follow-up, only 1 patient experienced chemoradiation because of merging ganglioneuroblastoma and receiving subtotal resection. No recurrence in other 5 cases [10].

Case reports

2016

A 12-year-old girl who was hospitalized due to neoplasm with spinal deformity in the right abdomen for 1 month. Based on a careful preoperative evaluation and found no obvious surgery contraindications, the patient was treated with surgical resection of the tumor and correction of the deformity by surgery. Postoperative pathologic examination confirmed it was a ganglioneuroma. After the operation, the patient recovered well. Her spinal deformity was corrected, and she was 5 cm taller. Complete resection of ganglioneuroma following with a low recurrence rate and a good prognosis, patient does not need further chemotherapy, radiation therapy, or other treatments. All follow-up radiographic studies demonstrated no relapse of the tumor in the following 18 months [11].

2015

Yilmaz et al. report an extremely rare case of lumbar spinal GN with conus medullaris invasion and extradural paraspinal extension. A 10-year-old girl presented with a history of worsening lower-back pain and an intermittent tingling sensation in the left leg. Neurological examination revealed reduced sensation in the left L2-L5 dermatomes. Magnetic resonance imaging revealed an intra- and extradural mass extending from the D11 to L5 vertebral body level. There was conus medullaris invasion by the tumor. After L1-L5 laminotomies, the patient underwent tumor resection.

Histopathological diagnosis was immature GN. [12].

2001

Bilateral and symmetric C1-C2 dumbbell ganglioneuromas producing severe spinal cord compression [13].

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