Case 1074

Paraspinal neuroblastoma with intraspinal extension

Published on 27.05.2001

DOI: 10.1594/EURORAD/CASE.1074
ISSN: 1563-4086
Section: Neuroradiology
Imaging Technique: MR
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Case Type: Clinical Cases
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Patient: 8 days, male

Clinical History:

An 8-day-old male patient was referred with intractable diarrhea, episodes of flushing, sweating, decreased motions of the lower extremities. A previous ultrasound examination of the patient revealed right-sided hydronephrosis, a paraspinal mass of the dorsal-lumbar area with intraspinal extension.

Imaging Findings:

An 8-day-old male patient was referred with intractable diarrhea, episodes of flushing, sweating, decreased motions of the lower extremities. A previous ultrasound examination of the patient revealed right-sided hydronephrosis, a paraspinal mass of the thoracic-lumbar area with intraspinal extension. Prior to the biopsy procedure which put the definite diagnosis, MRI study of the thoracic-lumbar spine with SE T1, FSE T2, post-gadolinium SE T1 sequences on sagittal, oblique coronal and axial planes was performed.

Discussion:

Neuroblastoma is a disease of infancy and childhood, seen once in every 7100 to 10000 births. The most affected children are in the age group of less than 5 year old. Neuroblastoma is the fourth most common tumor of the childhood. Males are slightly more affected. Neuroblastomas often originate in a paraspinal location, they can extend through the neural foramina to impinge on the thecal sac. One to four % of the patients present with spinal cord compression. Involvement of the spine occurs most frequently in thoracic and lumbar regions. Clinical findings vary according to the location and extent of disease. With spinal canal involvement the most common presenting symptoms include local pain and spinal cord dysfunction. The patient may have a paraspinal mass or signs of spinal cord compression such as impaired motor and sphincter function, weakness of the lower and upper extremities.

Cord compression is also common in terminal stages of the disease due to the frequent bone metastases. Cerebral and spinal cord parenchymal metastases from neuroblastoma are rare, however leptomeningeal metastases are more commonly seen. Neuroblastoma originates from primitive cells called neuroblasts, which are of neural crest origin. These neural crest cells embryologically form the adrenal medulla and the paravertebral sympathetic chain. The adrenal medulla and upper abdominal parasympathetic chain are the primary sites of 65 % of neuroblastomas, however the carotid ganglia, the aortic bodies, the organ of Zukerkandl may also be involved. Histopathologically neuroblastomas are composed of small round cells with hyperchromatic dense nuclei, these cells are primitive cells, which lack elements characteristic of further maturation. The tumor may be hemorrhagic, calcifications are not common. The imaging findings are erosion of the pedicle, widening of the foramina, scalloping of the vertebral body, widening of the spinal canal as the tumor extends through the neural foramina in direct roentgenograms. MRI demonstrates the paraspinal and intraspinal components of the tumor, as tumor extends through the intervertebral
foramina displacement and compression of the thecal sac and spinal cord can be clearly observed. The intraspinal component of tumor may spread through the epidural space over several levels, so resulting in cord block remote from the site of the paravertebral mass. The tumors rarely directly invade the intradural space. When the tumoral mass is an extension of the bony metastasis, irregular destruction of the vertebral body with extension of the soft tissue mass into the spinal canal is seen. The mass is relatively homogeneous and isointense to nervous tissue. Following the contrast administration the mass enhances heterogeneously. In the differential diagnosis of neuroblastomas, ganglioneuroma and ganglioneuroblastoma should be considered. Ganglioneuroma and ganglioneuroblastoma arise from the same cells as neuroblastoma. Ganglioneuroma is the most differentiated lesion and is composed almost entirely of mature ganglia cells. Ganglioneuroblastoma is a mixture of immature neuroblastoma and more mature cells. Ganglioneuroma and ganglioneuroblastoma tend to present later than neuroblastomas, usually seen in the 5-8 year-old age group. All tumors in the neuroblastoma, ganglioneuroblastoma, ganglioneuroma spectrum may have an identical appearance on imaging studies.

**Differential Diagnosis List:** Paraspinal neuroblastoma with intraspinal extension

**Final Diagnosis:** Paraspinal neuroblastoma with intraspinal extension

**References:**


Description: Sagittal FSE T2-weighted MR image shows a large extradural mass with isointense signal intensity to neural tissue at the level of thoracic-lumbar area, causing compression of the thecal sac and spinal cord. The spinal canal is widened smoothly. Origin:
**Description:** Coronal oblique FSE T2-weighted MR image shows the extension of large paraspinal mass through the widened neural foramina into the spinal canal, the paraspinal mass causes the compression of right kidney, leading to right-sided hydronephrosis. **Origin:**
**c**

**Description:** Axial SE T1-weighted MR image shows the paraspinal mass extending into the spinal canal through the widened neural foramen. **Origin:**

**d**

**Description:** Axial FSE T2-weighted MR image shows the paraspinal mass extending into the spinal canal through the widened neural foramen; note the right-sided hydrenephrosis. **Origin:**
Description: Sagittal post-gadolinium SE T1-weighted MR image shows the large extradural mass with homogeneous contrast enhancement, causing compression of the thecal sac and spinal cord. The spinal canal is widened smoothly. Origin:
**Description:** Coronal oblique post-gadolinium SE T1-weighted MR image shows the extension of large paraspinal mass through the widened neural foramina into the spinal canal, the mass has a homogeneous contrast enhancement, and causes the compression of right kidney, leading to right-sided hydrenephrosis. **Origin:**