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A case of Pelvic Ganglioneuroblastoma Totally Excised with Posteior-Sagittal and Abdominal Approach

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✔ Neuroblastoma usually presents as an upper abdominal mass arising from the adrenal gland. Recent experience with neuroblastoma of the spermatic cord, bladder and pelvis demonstrates the propensity of this tumour to arise in unusual areas. The total surgical excision of neuroblastoma cases are great important for prognosis. We report a case of pelvic ganglioneuroblastoma totally excised with posterior-sagittal and abdominal approach.

Key Words: Ganglioneuroblastoma, posterior-sagittal approach.

Neuroblastomas are the most common solid malignant tumours of infancy[1]. It is believed to originate from neural crest cells that normally give rise to the adrenal medulla and sympathetic ganglion[2]. Among these tumours 70 percent arise in the abdomen of which half are adrenal in origin[3]. Neuroblastomas have been reported predominantly to arise from paravertebral and prevertebral ganglia in the cervical, thoracic, lumbar and sacral regions and in postganglionic cells in the adrenal medulla[4].

The majority of pelvic neuroblastomas are believed to arise from the organ of Zukerkandl[4]. They are found in the presacral region and commonly present either as an asymptomatic palpable mass or with irritable bowel and urinary symptoms[2,5]. Pelvic neuroblastomas, although uncommon has been reported to have a more favourable prognosis than other more common sites of origin[4,5].

We present a case of ganglioneuroblastoma localised in pelvis and presacral region which was totally excised with posterior-sagittal and transperitoneal approach.

CASE REPORT

One year old boy was admitted to Ondokuz Mayıs University Children's Hospital Pediatric Surgery Department with acute urinary retention and with little and frequent urination and chronic constipation. On routine physical examination, a hard, fixed and solid lower abdominal mass was found. The fixed, hard mass with a diameter of 5x4 cm. was found between the sacrum and rectum in rectal examination.

Routine blood biochemistry was normal. Vanilmandelic acid is negative in spot urine. Carsinoembryogenic antigen, Alphafetoprotein levels and long bone radiograms were normal.

In ultrasonography bilateral pelvicaliectesia due to urinary bladder compression and a semisolid, smooth shaped, 66x38x42 mm. diameter mass with homogeneous density was revealed. In CT examination a smooth shaped presacral mass with dimensions of 5x5,5x4 cm, containing hypodense nodules was found (Figure 1).

We reached to pelvic mass with posterior-sagittal approach in the first stage of operation (Figure 2). Parasagittal muscle fi-

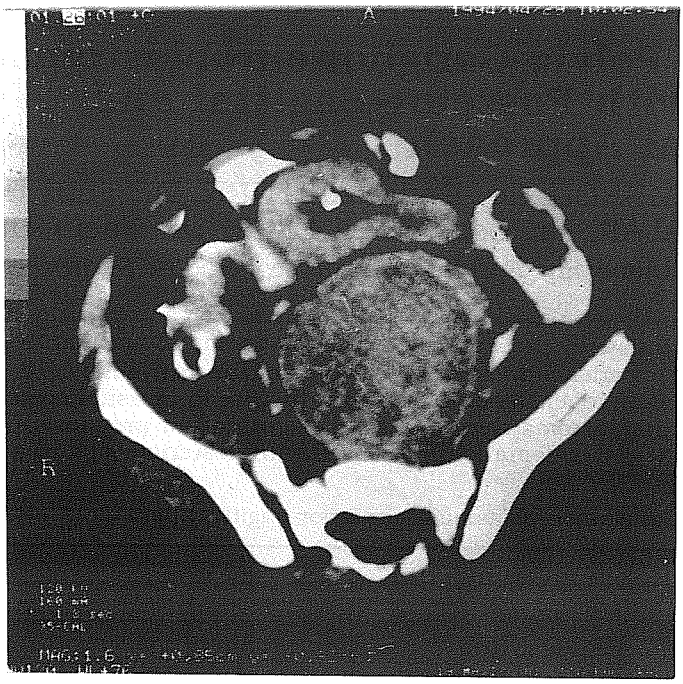

Figure-1: View of a 5x5,5x4,5 presacrally localised smooth shaped mass and containing nodulas in CT.

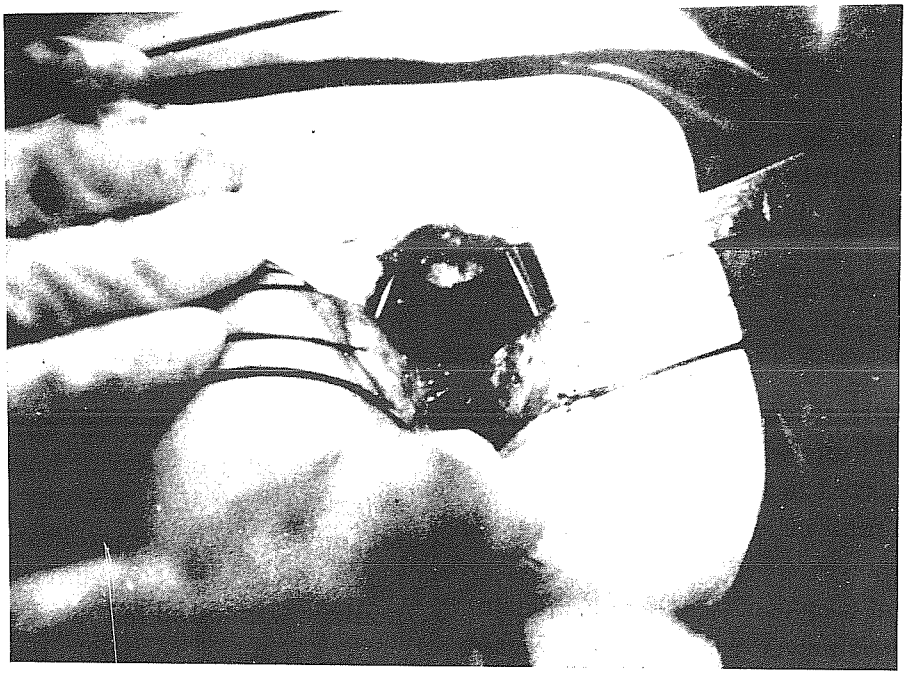

Figure-2: View of the presacral and intrapelvic mass in posterior–sagittal approach.

bers were separeted and muscle complex and levator ani muscle were divised. Mass was freed from posterior rectal wall and distal sacral region. Laparatomy was performed in the second stage of operation. The mass which was localised in presacral region, compressed the a. and v. iliaca and posterior rectal wall, urinary bladder and lower part of the left ureter. Then the tumor was totolly excised by the aid of the first stage.

Macroscopic examination revealed a 5x5x4 cm. diametered, capsulated, smooth shaped, semisolid pinkish-gray mass. Microscopicaly the tumor was composed of neuromal cells showing all stages of differentiation. The neuroblasts formed nests in which rosettes (Figure 3) and occasional mitosis were noted. The immature ganglion cells and bizarre multinucleated cells were scattered through the neuronal and scanty stromal elements (Figure 4). A pathologic diagnosis of differentiated neuroblastoma (Ganglioneuroblastoma) was made. Cisplatin 15 mg/m^2, Oncovin 1.5 mg/m^2 and Endoxan 300–400 mg/m^2 were started on postoperative 5th day. The patient was discharged on postoperative 12th day. Pediatric oncology department continued chemotherapy. After six months, the patient was well and no tumor relapse was found.

DISCUSSION

Pelvic sites of origin account for 2 to 4 percent of all neuroblastomas[4,6]. Most pelvic neuroblastomas present as either an asymptomatic palpable mass or with bowel or urinary symptoms[2,5]. Of 7 patients reported by Ghazali 3 presented with palpable sacral masses and 4 presented with acute urinary obstructive symptoms and an associated palpable mass[7]. Our patient presented with chronic constipation and acute uri-

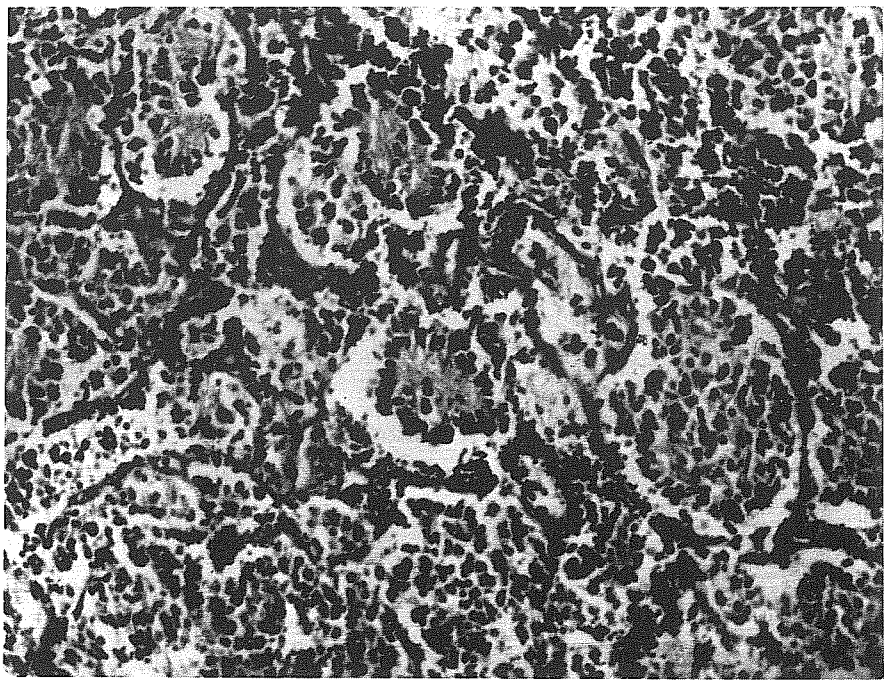

Figure-3: Microscopic appearence of neuronal rosettes. HE X 200.

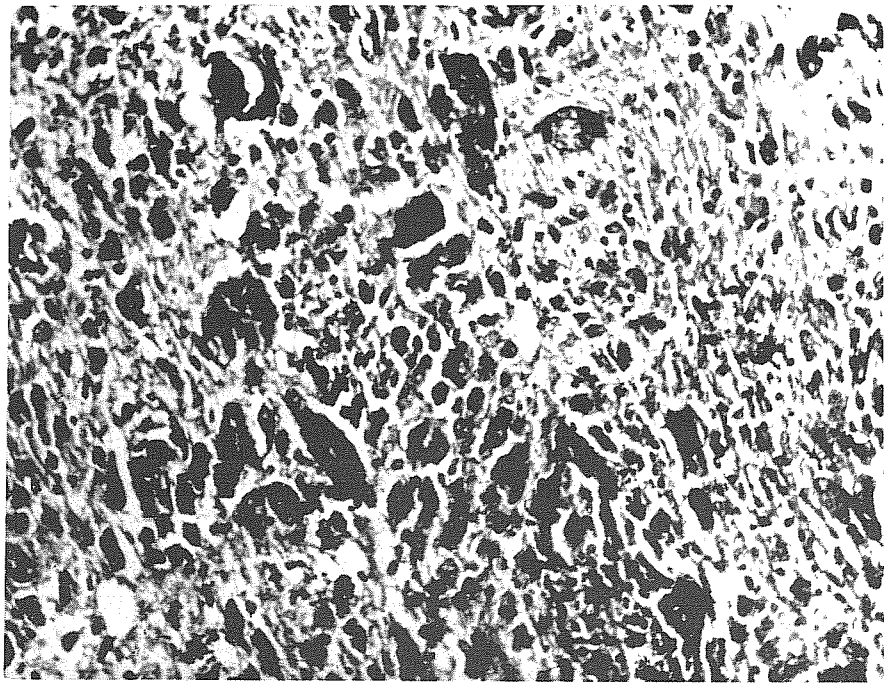

Figure-4: Microscopic appearence of cells and bizarre multinucleated cells. HE X 400.

nary retantion.

Histologically, neuroblastoma consist of a spectrum from ganglioneuroma, a well differentiated benign tumor, to the classical undifferentiated malignant form of neuroblastoma. Between the 2 extremes is the intermediate histological lesion, the ganglioneuroblastoma. Ganglioneuroblastomas shows features of the benign and malignant forms and may have an unpredictable clinical course with a reported 20 percent incidence of metastasis[8]. We did not detect metastasis of tumor in our patient. But we performed chemotherapy because the tumor has potential malignancy. Pelvic neuroblastomas presented mostly in children less than 2 years old[5,7]. Our patient was one year old.

Surgical approach of pelvic masses are extremely important. Abdominal and transsacral approach are applicable this aspect in pelvic masses[9]. Method of posterior-sagittal approach as in our case was not found in the literature for pelvic and presacral tumors. Transsacral approach may cause neurological damage[9]. It is well known that the risk of neurological damage is lower in posterior-sagittal approach. Posteior-sagittal approach gives better exposure then transsacral approach. So we used this surgical technique. We performed posteror-sagittal incision in first stage. Mass was freed from posterior rectal wall and anterior space of sacrum with this approach. In stage two, transperitoneal approach was performed on supine position and the mass was removed totally. The total surgical excision of neuroblastoma cases are great important for prognosis(4). Posterior-sagittal approach may be used for rectal atresia, rectal stenosis, persistent rectourethral or rectovaginal fistulae, urethral diverticulum and rectal trauma[10].

We beleive in pelvic masses, such as neu-

roblastoma posterior-sagittal approach in the first stage could allow the excision of tumor totally. For these reason we propose this surgical technique in pelvic neuroblastoma.

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