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Case Report

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Primary neonatal sacrococcygeal neuroblastoma masquerading as a teratoma: A case report

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KEYWORDS

Primary, Sacrococcygeal, Neuroblastoma, Neonatal

ABSTRACT

Background: Neonatal tumors comprise about 2% of all pediatric malignancies, with neuroblastoma having the highest incidence. Neuroblastoma involving the adrenal medulla and sympathetic ganglia is the most typical scenario in infancy, while the pelvic variant is rare. We report this case because of the unusual and rare presentation of neuroblastoma in a newborn baby mimicking sacrococcygeal teratoma.

Case Presentation: A newborn male baby presented with a firm sacral mass, about 5 x 7 cm, with normal overlying skin. MRI revealed an intrapelvic mass extending to the sacral region encasing the coccyx. After preparation, complete tumor excision was performed, and the diagnosis of neuroblastoma was confirmed by histological and immunohistochemical study.

Conclusion: Primary neonatal neuroblastoma presenting as a sacrococcygeal mass is a rare and atypical clinical finding of neuroblastoma. It is hard to diagnose this sort of tumor preoperatively unless the mass is subjected to histological and immunohistochemical analysis after tumor excision.

INTRODUCTION

Neonatal tumors comprise about 2% of all pediatric malignancies, with neuroblastoma and teratoma having the highest incidence of around 28 to 39% in this age group.[1] Neuroblastoma is an embryonal tumor arising from neural crest cells that give rise to the sympathetic ganglia and adrenal medulla.[2,3] It most commonly arises in the abdomen in (65%) of cases, with more than half originating from the adrenal gland.[4] Other sites of involvement are the neck, chest, and rarely the pelvis, with an incidence of 3.4%.[5] We report this case because of the atypical and rare presentation of neuroblastoma in a newborn baby that clinically mimics Altman type III sacrococcygeal teratoma

CASE REPORT

A full-term male baby was delivered by normal vaginal delivery. On routine neonatal examination, a swelling was noticed in the sacrococcygeal region. The swelling was firm and round, 5 x 7 cm in size, in the sacral region with normal overlying skin (Fig. 1). The abdomen was not distended, and no mass was palpable. This mass was not detected on antenatal ultrasonography. After clinical evaluation, Serum alpha-fetoprotein was 401ng/ml. Urinary vanillylmandelic acid and Homovanillic acid were not performed preoperatively. Magnetic resonance imaging revealed a presacral mass extended downward and encasing the fifth sacral vertebra and the coccyx (Fig. 2A, 2B). Based on the clinical and radiological findings, the diagnosis of sacrococcygeal teratoma was made.

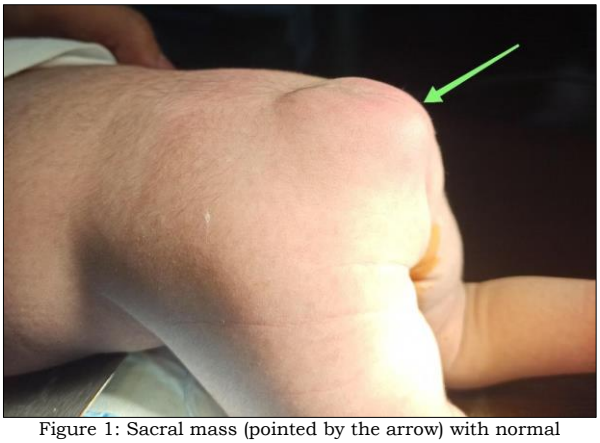

Figure 1: Sacral mass (pointed by the arrow) with normal overlying skin

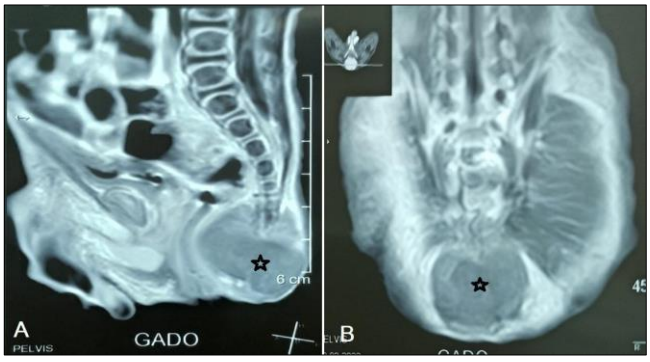

Figure 2 (A and B): MRI with GADO (in both sagittal and coronal section indicated by an asterisk) showing presacral mass extended downward and encasing the fifth sacral vertebra with the coccyx.

After preoperative preparation, the surgical procedure was performed at the age of five days under general anesthesia in a prone position with a pillow under the pelvis, and through a chevron incision, complete tumor excision including the coccyx with a safety margin was performed after ligation of the feeding vessel (Fig. 3A, 3B). The tissue sample was sent for histological and immunohistochemical study, which gave the diagnosis of neuroblastoma (Fig. 4a, 4b, 4c). Postoperatively, urinary vanillylmandelic acid and homovanillic acid were checked monthly for six months and the results were normal. Magnetic resonance imaging was also repeated at the third and sixth months postoperatively, which showed a clear pelvic and sacral region without any evidence of local recurrence or distant metastasis. Now the patient is six months old, and has active limbs, without any evidence of mass lesion at the site of operation.

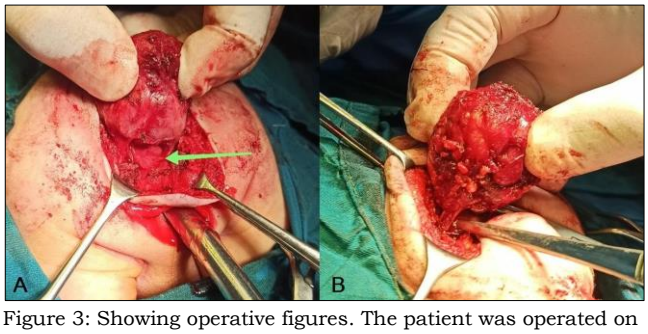

Figure 3: Showing operative figures. The patient was operated on in the prone position by a Chevron incision. A) During mobilization of the tumor with preservation of the rectum (marked by the arrow). B) Complete isolation of tumor with an intact capsule, the feeding vessels marked by forceps

Pathological examination: Microscopically, sections show malignant tumors composed of variably sized nodules separated by delicate fibrovascular septae. The individual nodule consists of sheets of rather monomorphic small rounded cells having rounded deeply stained nuclei with invisible cytoplasm, set in a background of light eosinophilic fibrillary material. Focal pseudorosettes were noticed (Fig. 4a). The coccyx was free of the tumor. The histologic findings were that of malignant small round cell tumor in favor of undifferentiated neuroblastoma of unfavorable histologic type) with a mitosis-karyorrhexis index (MKI) of (<2%). The biopsy was reviewed by 2 independent pathologists. The tumor was categorized as (stage I) according to the international neuroblastoma staging system (localized, completely excised tumor with free margin confirmed by pathological examination, with no distant metastasis).

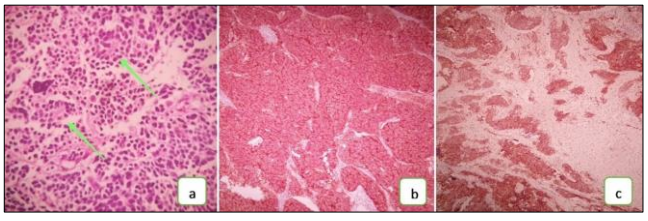

Figure 4: a) Hematoxylin and eosin, power 10x showing proliferating small cells with scanty cytoplasm in the cobwebby fibrillary background with pseudorossette formation (arrow). b) Immunohistochemistry, showing diffuse positivity for CD56. c) Immunohistochemistry, showing diffuse positivity for chromogranin

Immunohistochemical examination: A panel of markers was done, and the tumor cells showed strong diffuse reactivity for CD56, a marker of neural/neuroendocrine tissue (membranous positivity), chromogranin A (cytoplasmic positivity), & synaptophysin (cytoplasmic positivity) (Fig. 4b, 4c). They were negative for AE1/3, desmin, SMA, CD45, CD99, CD117, and WT-1 (thus excluding teratoma, Ewing's sarcoma, lymphoma, Wilms' tumor, and sarcoma). Genetic tests were not performed (not available in our locality).

DISCUSSION

German physician Rudolf Virchow first described neuroblastoma in 1864. James Homer Wright coined this term in 1910, stating that neuroblastoma arises from the neural crest cells.[6] Congenital neuroblastoma is referred to as neuroblastoma presented in the first month of life and it constitutes about 1% of all neuroblastoma presenting in other age groups.[7,8] Neuroblastoma commonly arises from the adrenal gland, but it can also originate from different anatomic locations of the sympathetic chain, such as the neck, chest, abdomen, and pelvis. Pelvic neuroblastoma occurs in about 5% of all neuroblastoma cases and comprises about 0.25% of all neonatal malignancies.[9,10] Boys are slightly more affected than girls, with a male-to-female ratio of 1.2:1.[11] Several environmental causative factors are implicated in the development of neuroblastoma, including maternal exposure to alcohol, phenobarbital, pesticides, or exposure to electromagnetic fields.[12] A few cases of sacrococcygeal neuroblastoma were reported in the literature with different ages at presentation. In 2005, Tanaka et al. reported a case of neonatal neuroblastoma similar to sacrococcygeal teratoma

(Altman type III).[13] In 2008, Watanabe et al. reported a case of sizeable presacral neuroblastoma in a two-month-old girl.[14]

Fatih et al. in 2013 also reported a case of sacral neuroblastoma in a four-month-old boy.[10] Prenatal diagnosis of congenital neuroblastoma is possible after 32 weeks of gestation and approximately 93% of the antenatally diagnosed tumors were of adrenal origin.[12] The clinical presentation of sacrococcygeal neuroblastoma may include abdominal distention and urinary retention if the intrapelvic component of the tumor is big in size. However; our patient was asymptomatic apart from the sacral mass, which was detected during routine neonatal evaluation. The diagnosis of sacrococcygeal neuroblastoma is hard to make before surgical resection and pathological examination. In some studies, the preoperative urinary vanillylmandelic acid and Homovanillic acid are found to be normal or slightly elevated in such cases.[13,14] In our case, urinary vanillylmandelic acid and Homovanillic acid were not performed preoperatively, instead, we did serum alpha-fetoprotein because the preoperative diagnosis was teratoma. The possible differential diagnosis could be either benign or malignant tumors. Benign tumors are lipoma, hemangioma, meningocele, cystic lymphangiomas, and teratoma, while malignant tumors are cystic neuroblastoma, teratoid Wilms tumor, and ependymoblastoma.[15]

Complete surgical excision with tumor-free margins is the goal of the treatment. In our patient, the good postoperative clinical well-being despite the unfavorable type of tumor may be due to complete tumor excision with free margin without any local residual, and this was performed at an early age (5 days old); in addition, the distant metastasis was not recorded at the initial MRI study nor on the follow-up visits.

To conclude, primary neonatal neuroblastoma affecting the sacrococcygeal region is a rare clinical scenario and could be misdiagnosed as a sacrococcygeal teratoma on physical examination and radiology. Histopathological and immunohisto-chemical studies of surgically excised tumor tissue thus form a gold standard for definitive diagnosis and further clinical management of the patient.

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