Case Report

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Lipoblastoma-Neuroblastoma: A Rare Paediatric Presentation

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Abstract

Blastomas are tumors that are unique to childhood. They are thought to arise from immature, primitive tissues that present persistent embryonal elements on histology, affect a younger paediatric population and are usually malignant. A blastoma is thought to be a genetic error of normal development rather than the result of any external, environmental factor [1, 2].

Keywords: Lipoblastoma; Neuroblastoma; Synchronous Presentation; Paediatric.

Case Report

An 8 month female presented with gradually increasing swellings in bilateral inguinal regions (left>right) since one month. The child had no other complaints and was thriving well. On local examination, a 3x3 cm well defined, non-tender, non-reducible, mobile mass in the left inguinal region with no cough impulse and a similar smaller mass was present in the right inguinal region. The external genitalia were normal with a soft abdomen and no organomegaly. An ultrasonography showed hypoechoic mass in left inguinal region suggestive of a lipoma or irreducible omental hernia. Amass was visualized in the retroperitoneum with sub-centimetric abdominal lymphadenopathy. Computed Tomography revealed 3.6 x 2.3 x 2 cm mass in retroperitoneum compressing the IVC antero-medially and a2.4x2.3x1.8 cm lesion in the left inguinal region. Tumor markers were negative (AFP was 14.43 ng/ml, B HCG < 0.13 mIU/ml, CA 125 was 22.5 U/ml, 24hour VMA-). An excisional biopsy of both inguinal swellings was suggestive of lipoblastomatosis, positive for S100P and p16. The retroperitoneal mass was also excised. The histology was suggestive ofa ganglioneuroblastoma with focal areas reminiscent of lipoblastoma/lipoblastomatosis positive for synaptophysin and chromogranin. PET scan did not show any uptake or evidence of tumor activity. The child was not given any chemotherapy, is doing well and is being followed closely.

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Discussion

The suffix 'blastoma' reflects a presumed relationship to primitive, 'blastic' elements and a dysontogenetic character. The pediatric embryonal tumors are presumed to arise from immature cellular populations that have not completed the process of differentiation to mature tissue phenotype during pre and postnatal development and are diagnosed in the majority during the first 5 years of life [1,2,3].

Neuroblastoma is a common paediatric malignancy and along with the other blastomas (nephroblastoma, hepatoblastoma, medulloblastoma) accounts for nearly 25% of solid tumors in the paediatric age group. The rare types of blastomas (lipoblastoma, osteoblastoma, chondroblastoma, hemangioblastoma, gonadoblastoma, sialoblastoma, pleuropulmonary blastoma, pancreatoblastoma, pineoblastoma, and medullomyoblastoma) individually account for less than 1% of paediatric malignancies; collectively they may be responsible for upto 5% of

paediatric tumors.

Lipoblastomas are rare benign adipose tumors arising from immature adipocytes demonstrating varying degrees of differentiation [5,6]. Lipoblastomas show male predominance and are found almost exclusively in children younger than 5 years [4, 5, 6]. There are two forms of lipoblastoma: (a) the lipoblastoma – a superficial, well circumscribed, and encapsulated lesion, which macroscopically resembles a lipoma; (b) the lipoblastomatosis – a deeply located, poorly circumscribed, macroscopically diffuse and infiltrative lesion. Lipoblastomatosis is commoner; affecting 66% of all cases [7], and has a tendency to recur post-surgically.

Lipoblastomas present as soft, non-tender, lobular masses of variable size, which enlarge rapidly.These tumors show a peculiar predilection for the left side of the body, which may suggest an association with genes of asymmetry, and a breakpoint on chromosome 8q 11–13 [4]. Treatment of lipoblastomas is complete surgical excision, with recurrence rates that range from 9% to 22%, usually associated with incomplete excision [6].

Neuroblastoma is the most common extracranial solid tumor of infancy. It is an embryonal malignancy of the sympathetic nervous system arising from neuroblasts which commonly populate the sympathetic ganglia, adrenal medulla, and other sites. The pattern of distribution of these cells correlates with the sites of primary neuroblastoma presentation.Origin and migration pattern of neuroblasts during fetal development explains the multiple anatomic sites where these tumors occur; location of tumors varies with age. Tumors can develop in the abdominal cavity (40% adrenal, 25% paraspinal ganglia) or other sites (15% thoracic, 5% pelvic, 3% cervical tumors, 12% miscellaneous). Ganglioneuromas, ganglioneuroblastomas, and neuroblastomas differ from one another in terms of the stage of neuroblast maturation. The ganglioneuromas, which are composed of mature ganglion cells, are considered benign tumors. Ganglioneuroblastomas are less mature forms. They are regarded as more aggressive tumors, and they generally develop in small children

The occurrence of simultaneous embryonal tumors from different origins is very rare. They are commonly associated with hereditary syndromes. A concurrent neuroblastoma and adrenocortical tumor in a patient with Beckwith-Wiedemann syndrome has been reported. Another report of a congenital pineoblastoma and concurrent parameningeal embryonal rhabdomyosarcoma in early infancy is the first of its kind.

The association between adipose tissue and neural tissue has been in the form of fatty replacement of ganlioneuromas. This occurrence is very rare and only one case of lipomatous retroperitoneal ganglioneuroma and three cases of mediastinal ganglioneuroma with fatty replacement are reported in the literature [10].

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