Massive paraganglioma in an infant

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ABSTRACT

We report here a 7-month-old baby presenting with a large paraganglioma arising from the right side of neck and extending to the scalp. Catecholamine screening was within normal limits. Ultrasound of the abdomen, CT-scan, and whole body MRI excluded any other coexisting neuroendocrine tumor. The tumor was treated by radical excision, and the resultant soft tissue defect was resurfaced using a pectoralis major muscle flap with split skin grafting. There has been no recurrence or metastasis during the 2-year follow up. To the best of our knowledge, this is the youngest reported patient with paraganglioma of the neck. Surgical excision of paraganglioma is feasible and curative in the absence of metastasis.

KEY WORDS: Neuroendocrine tumor, Paraganglioma

Paragangliomas are rare neuroendocrine tumors capable of secreting a variety of hormonal substances; but they rarely elicit clinically apparent endocrine or metabolic effects.[1] We report here a 7-month-old infant presenting with massive paraganglioma of neck and scalp, unique in several aspects; first, to the best of our knowledge, this is the youngest reported patient with this rare tumor. Second the tumor was very large; however radical excision was curative.

CASE REPORT

A 7-month-old infant was admitted with a large mass on the right side of neck and scalp [Figure 1]. The mass had gradually increased in size over 5 months, and fungation through skin occurred 15 days before admission. Family history was not contributory. The swelling displaced the right ear anteriorly and superiorly. The mass was firm and rubbery on palpation with restricted mobility over the scalp. Investigations included hematocrit (hemoglobin 10 mg/dl) and biochemical screening for catecholamines, which were normal. Chest radiograph, ultrasound of abdomen, and MRI of the whole body was done to detect any coexisting tumor or distant metastases, and none were present. The child was prepared for surgery and radical excision of the tumor was performed under general anesthesia, and the tumor was easily separated from the skull and removed along with the overlying skin. It was in close proximity to the carotid sheath but not infiltrating any major blood vessels or nerves. Complete excision was possible and the resultant defect was closed with a right-sided pectoralis major muscle flap with split skin grafting. Postoperative recovery was uneventful.

Histopathological examination of the tumor revealed a highly cellular field with atypically proliferated round or ovoid cells disposed in solid masses, sheets, or groups surrounded by thin fibrous septa containing small, thin-

walled blood vessels. The cells showed deeply stained round nuclei and strongly argyrophilic cytoplasmic granules stained by Grimelius stain. There was mild anisonucleosis and nuclear pleomorphism but no mitotic figures. Immunohistochemistry was positive for the neuroendocrine tumor markers PGP 9.5, neuron-specific enolase [Figure 2], and S-100 (neuronal protein). The pathological diagnosis was paraganglioma. The child is under regular follow up and there is no recurrence or metastasis till date.

DISCUSSION

Paragangliomas of the head and neck are a group of rare, usually benign, slow-growing tumors developing from the extra adrenal paraganglionic tissue,\(^1\) chiefly the carotid body, jugular bulb, tympanic plexus, vagal body, aorticopulmonary paraganglia, sinoatrial chambers, orbit, and larynx.\(^2\)–\(^7\) The usual age of presentation is after 40 years; the youngest patients previously described were over 12 years of age.\(^8\),\(^9\) No definite sex predilection has been described in the literature. The clinical behavior of paraganglioma is determined by the cellular characteristics, secreting capabilities, and tumor location.\(^2\) Surgery is the mainstay of treatment, to be supplemented by radiotherapy,\(^2\)–\(^4\) if necessary. The selection of treatment depends on the size, location, and biologic activity of the tumor, as well as the overall fitness of the patient.\(^5\) Malignancy is rarely seen in paraganglioma and is defined by the existence of metastasis rather than histology.\(^2\) Recurrence after complete excision is rarely seen with reported recurrence rates varying between 0\(^5\) and 15.9%.\(^3\) Our patient is unique in several aspects. First, to the best of our knowledge, he is the youngest reported patient with this rare tumor. There was no clinical or biochemical evidence of catecholamine over activity. Second, the tumor was very large; however, it was amenable to radical surgical excision. Such a large paraganglioma of head and neck (1500 g) has not been reported previously in an infant. The soft tissue defect after excision of the tumor and involved skin required pectoralis major muscle flap reconstruction. Split skin grafting was used to resurface the defect. The pathological diagnosis was confirmed by immunohistochemistry, the tumor cells were positive for neuroendocrine tumor markers such as PGP 9.5, neuron-specific enolase,\(^6\) and S-100 (neuronal protein).\(^6\),\(^10\) Postoperative recovery of the baby was good and there is no evidence of local recurrence or distant metastasis during follow up. We conclude that surgical excision of paraganglioma is feasible and in the absence of metastasis it is curative.

REFERENCES