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Life-threatening Airway Obstruction: An Unusual Presentation of a Cervical Mass During Infancy

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Abstract Cervical masses during infancy are almost always benign. The more common differential diagnoses include congenital malformations of the vascular or lymphatic systems, remnants of the branchial arches, sternocleidomastoid tumour of infancy, and lymphadenitis. Benign tumours such as dermoid cysts or teratomas are uncommon, and malignant lesions are extremely rare. The authors reported a 4-month-old infant who presented with respiratory arrest secondary to upper airway obstruction caused by a cervical neuroblastoma. The child was successfully resuscitated but required mechanical ventilation. Subsequent surgical excision without chemotherapy led to complete clinical, radiological, and biochemical remission. Neck masses during early childhood should be handled with care and malignant lesions should be distinguished from the more common but self-limiting causes.

Key words Airway obstruction; Congenital muscular torticollis; Head and neck neoplasms; Horner syndrome; Neuroblastoma

Introduction

Neck masses are common paediatric problems with a wide variety of pathologies.[1,2] During infancy, the great majority of cases are due to malformations of the head and neck structure or congenital muscular torticollis (sternocleidomastoid tumours of infancy).[3,4] The conditions are usually benign and most are recognisable on clinical grounds. However, malignant conditions, though rare, may mimick these lesions. The following case report is to remind of the need for careful assessment and follow-up in any infant who presents with an apparently benign mass in the cervical region. The differential diagnoses of infantile neck masses will be presented, followed by a discussion of the management of cervical neuroblastoma in young children.

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Case Report

A 4-month-old baby girl was referred to our hospital for a gradually enlarging mass on the right side of the neck. She was born prematurely at 33 weeks of gestation from a pregnancy that was complicated by severe pre-eclampsia. The birth weight was 1.57 kg. The stay in the Special Care Baby Unit was uneventful and she was discharged at one month of age.

Her parents noticed a nodule over the right side of the neck when she was two months old. There was no abnormal head posture and the baby appeared well and thriving. The mass was thought to be a sign of congenital muscular torticollis and the child was treated with physiotherapy. However, the mass grew gradually in size and the baby subsequently developed noisy breathing and feeding difficulty. An ultrasonography showed a solid right

parapharyngeal mass with calcification and hence the child was referred.

On admission, the child was noted for a right-sided neck swelling and an inspiratory stridor. She remained playful and there was no choking on feeding. The respiratory rate was 28 per minutes and the air entry was satisfactory. The transcutaneous oxygen saturation was 99% in room air. There were no lymphadenopathy, Horner's syndrome, heterochromia iridis, hepatomegaly, or any abdominal mass found. While further investigations were organised, she suddenly lapsed into respiratory arrest twelve hours after admission. She was immediately resuscitated and spontaneous respiration resumed after manual bagging and oropharyngeal suction. However, she lapsed into respiratory distress and cyanosis an hour later, and was then intubated with a 4-mm endotracheal tube for mechanical ventilation and intensive care. Subsequent CT showed a heterogeneous, contrast-enhancing mass in the right parapharyngeal region that measured 5.5 × 3.3 × 5.4 cm, extending from orophargnx to the thoracic inlet (Figure 1). The lesion crossed the midline and displaced the upper airway, jugular vein and carotid arteries.

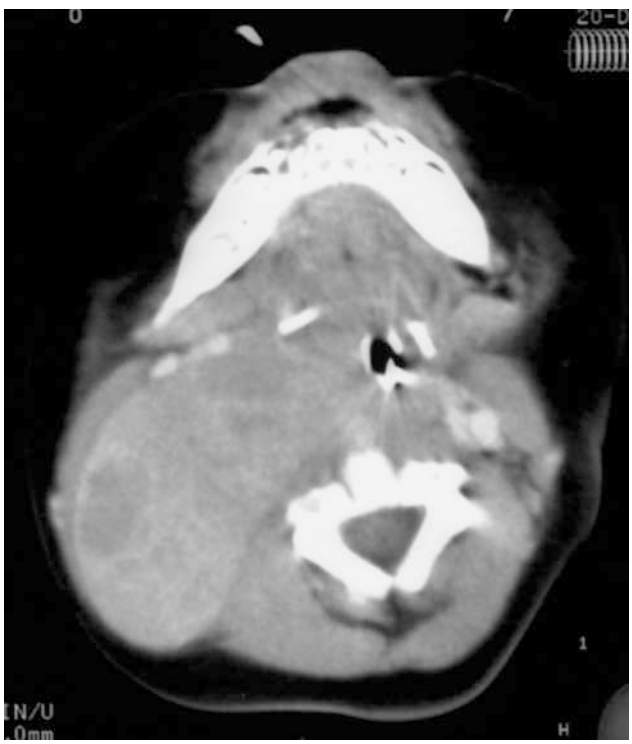

Figure 1 CT scan of the neck showing a large mass in the right parapharynx with heterogeneous contrast enhancement displacing the trachea to the left and the vascular sheet in the front.

An immediate surgery was undertaken to remove the mass completely. Intraoperatively, a huge soft tissue mass of 6 cm in maximum dimension arising from the prevertebral fascia on the right side of the neck was removed (Figure 2). The diagnosis of neuroblastoma was confirmed on histopathology. The patient made an uneventful postoperative recovery, and was weaned from the ventilator the next day after surgery. Subsequent investigations including CT of abdomen, bone scintigraphy, and meta-iodobenzylguanidine imaging were negative. The urinary levels of vanillylmandelic acid and homovanillic acid were elevated initially but were normalised on subsequent measurements. The child remained in clinical, radiological, and biochemical remission at 2 years of age.

Discussion

Masses at the cervical region are a common finding in early infancy.[1-3] The great majority of these lesions are benign (Table 1) and may represent malformations of the branchial arches, vascular or lymphatic system, or other cervicothoracic structures.[5-7] Other differential diagnoses include congenital muscular torticollis, infective conditions, or benign neoplasms.[4,8-12] Malignancies are rarely reported.[13-15] Most of these benign pathologies can be diagnosed on clinical grounds alone, but their relative frequencies have not been systematically studied. Imaging may be indicated when the anatomy or the consistency of the lesion has to be better defined.[1,3]

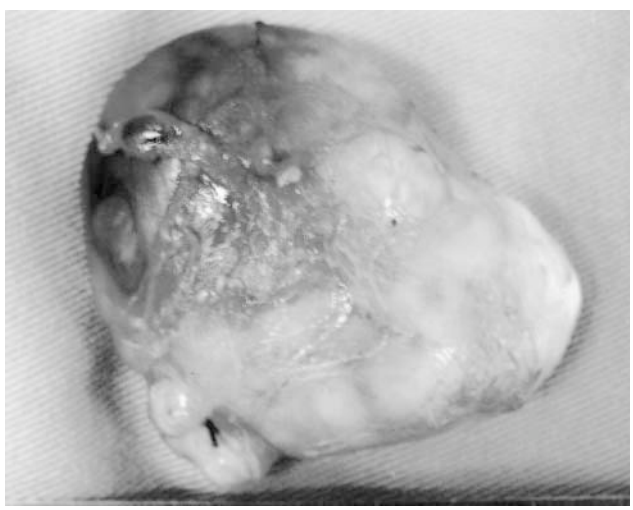

Figure 2 Gross appearance of the resected mass measuring 6 cm in its maximum dimension.

Table 1 Differential diagnosis of neck masses during infancy[1-3]

Congenital anomalies
Branchial cysts [5]
Thyroglossal cysts [7]
Ectopic thymus and thymic cysts [6]
Vascular or lymphatic anomalies
Haemangiomas
Cystic hygromas
Inflammatory conditions
Sternocleidomastoid tumour (congenital muscular torticollis)
Ranula
Lymphadenitis and neck abscesses [8]
Benign neoplasms
Dermoid cysts and teratomas [9,10]
Lipoblastomas [11]
Myofibromas [12]
Malignancies
Neuroblastomas
Germ cell tumours [13]
Rhabdomyosarcomas [14]
Lymphomas

Pseudotumours associated with congenital muscular torticollis are common neck masses found in young infants, affecting 0.3-1.9% of young children. A local tertiary referral center had recruited 1,086 infants during a 12-year period.[4] Given its common occurrence, it is not surprising that an initial diagnosis of congenital muscular torticollis was made in the present case. Congenital muscular torticollis usually occurs in babies delivered at term and become noticeable some weeks after birth. Over 90% of congenital muscular torticollis present in the first three months of life. They may occur on either side of the neck, involving the middle or lower third of the sternocleidomastoid muscle, and present as palpable swellings along the muscle. The majority of babies also present with tilting of the head to the opposite side or facial asymmetry. The diagnosis is made clinically, and physical treatment leads to resolution in most instances.

In contrast, malignant lesions at the neck are extremely rare in early infancy.[2] Neuroblastoma, germ cell tumour, lymphoma, and rhabdomyosarcoma may present as a neck mass.[1,3] Cervical and cervicothoracic neuroblastomas are probably the commonest reported lesion in young infants, although they account for only 3-5% of all neuroblastomas.[15,16] Originating from the sympathetic nervous tissue behind the vascular sheet, neuroblastomas of the neck often present with obstruction of the upper aerodigestive tract and cause stridor or feeding difficulty,[17] suggesting that most cases are not recognised in the early stage. Unilateral Horner's syndrome and heterochromia are other manifestations,[18,19] but they may be difficult to notice prior to operation when circulating levels of catecholamines are elevated. Recurrent laryngeal nerve or phrenic nerve palsy may happen if the tumour extends to the thoracic inlet.[20] The presence of intralesional calcifications and raised levels of urinary catecholamines are important clues to the diagnosis. However, misdiagnosis as cystic hygroma or infective adenitis has been reported.[18,21]

Primary cervical neuroblastomas used to portend a poor prognosis in earlier series. All of the four children treated in a single institution from 1944-1968 died from the disease despite aggressive treatment.[22] In the modern era, cervical neuroblastomas are a highly curable disease[17,19,23] in contrast to classical forms of neuroblastomas seen in older children. Primary adrenal neuroblastomas usually present beyond infancy and are often disseminated at the time of first presentation.[24] Despite aggressive treatment with a combination of surgery, chemotherapy, radiotherapy, and megatherapy followed by autologous haematopoetic stem cell transplantation, only one-third of patients can survive into the long term.[25] Cervical neuroblastomas, on the other hand, often present with localised disease[17,18] and are negative for N-myc gene amplification when tested,[18] a favourable prognostic factor in localised neuroblastomas.[26] Among 34 children with cervical or cervicothoracic neuroblastoma, 81% remained in first complete remission after treatment.[17] In a smaller series of 11 patients, all but one patient survived after operation.[18] The diagnosis of cervical neuroblastoma was only made at postmortem in the fatal case. For resectable tumours, primary surgery alone is often curative and complete excision may not be necessary for long-term survival.[27] An occasional child who relapses after incomplete resection may still be salvageable with combination chemotherapy.[28] Radiation therapy should be avoided. Cushing et al[23] reported two cases of thyroid carcinoma among five survivors of early childhood cervical neuroblastoma who had been treated with radiotherapy. For more extensive disease, neo-adjuvant chemotherapy followed by delayed surgery may be equally effective.[17,19,20]

In summary, neuroblastoma and other malignancies are rarely found in the cervical region of young infants. Care must be taken to distinguish them from the more common but benign infantile neck masses. Timely diagnosis and treatment is often rewarding for this rare but life-threatening malignancy.

References

1. Castellote A, Vazquez E, Vera J, et al. Cervicothoracic lesions in infants and children. Radiographics 1999;19:583-600.
2. Davenport M. ABC of general surgery in children. Lumps and swellings of the head and neck. BMJ 1996;312:368-71.
3. Turkington JR, Paterson A, Sweeney LE, Thornbury GD. Neck masses in children. Br J Radiol 2005;78:75-85.
4. Cheng JC, Tang SP, Chen TM, Wong MW, Wong EM. The clinical presentation and outcome of treatment of congenital muscular torticollis in infants--a study of 1,086 cases. J Pediatr Surg 2000;35:1091-6.
5. Rea PA, Hartley BE, Bailey CM. Third and fourth branchial pouch anomalies. J Laryngol Otol 2004;118:19-24.
6. Khariwala SS, Nicollas R, Triglia JM, et al. Cervical presentations of thymic anomalies in children. Int J Pediatr Otorhinolaryngol 2004;68:909-14.
7. Lindstrom DR, Conley SF, Arvedson JC, Beecher RB, Carr MH. Anterior lingual thyroglossal cyst: antenatal diagnosis, management, and long-term outcome. Int J Pediatr Otorhinolaryngol 2003;67:1031-4.
8. Leung AK, Robson WL. Childhood cervical lymphadenopathy. J Pediatr Health Care 2004;18:3-7.
9. Pryor SG, Lewis JE, Weaver AL, Orvidas LJ. Pediatric dermoid cysts of the head and neck. Otolaryngol Head Neck Surg 2005; 132:938-42.
10. Hasiotou M, Vakaki M, Pitsoulakis G, et al. Congenital cervical teratomas. Int J Pediatr Otorhinolaryngol 2004;68:1133-9.
11. Rosen A, Jedynak AR, Respler D. Lipoblastoma of the neck mimicking cystic hygroma. Otolaryngol Head Neck Surg 2005; 132:511-3.
12. Beck JC, Devaney KO, Weatherly RA, Koopmann CF Jr, Lesperance MM. Pediatric myofibromatosis of the head and neck. Arch Otolaryngol Head Neck Surg 1999;125:39-44.
13. Touran T, Applebaum H, Frost DB, Richardson R, Taber P, Rowland J. Congenital metastatic cervical teratoma: diagnostic and management considerations. J Pediatr Surg 1989;24:21-3.
14. Furze AD, Lehman DA, Roy S. Rhabdomyosarcoma presenting as an anterior neck mass and possible thyroid malignancy in a seven-month-old. Int J Pediatr Otorhinolaryngol 2005;69: 267-70.
15. Moukheiber AK, Nicollas R, Roman S, Coze C, Triglia JM. Primary pediatric neuroblastic tumors of the neck. Int J Pediatr Otorhinolaryngol 2001;60:155-61.
16. Haase GM. Head and neck neuroblastoma. Semin Pediatr Surg 1994;3:194-202.
17. Haddad M, Triglia JM, Helardot P, et al. Localized cervical neuroblastoma: prevention of surgical complications. Int J Pediatr Otorhinolaryngol 2003;67:1361-7.
18. Abramson SJ, Berdon WE, Ruzal-Shapiro C, Stolar C, Garvin J. Cervical neuroblastoma in eleven infants--a tumor with favorable prognosis. Clinical and radiologic (US, CT, MRI) findings. Pediatr Radiol 1993;23:253-7.
19. Ogita S, Tokiwa K, Takahashi T, Imashuku S, Sawada T. Congenital cervical neuroblastoma associated with Horner syndrome. J Pediatr Surg 1988;23:991-2.
20. Kapoor V, Lodha R, Agarwala S. Superior mediastinal syndrome with Rowland-Payne syndrome: an unusual presentation of cervico-mediastinal neuroblastoma. Pediatr Blood Cancer 2005; 44:280-2.
21. Ganick DJ, Kodroff MB, Marrow HG, Holbrook CT, Pories WJ. Thoracic neuroblastoma presenting as a cystic hygroma. Arch Dis Child 1988;63:1270-1.
22. Brown RJ, Szymula NJ, Lore JM Jr. Neuroblastoma of the head and neck. Arch Otolaryngol 1978;104:395-8.
23. Cushing BA, Slovis TL, Philippart AI, et al. A rational approach to cervical neuroblastoma. Cancer 1982;50:785-7.
24. Schwab M, Westermann F, Hero B, Berthold F. Neuroblastoma: biology and molecular and chromosomal pathology. Lancet Oncol 2003;4:472-80.
25. Matthay KK, Villablanca JG, Seeger RC, et al. Treatment of high-risk neuroblastoma with intensive chemotherapy, radiotherapy, autologous bone marrow transplantation, and 13-cis-retinoic acid. Children's Cancer Group. N Engl J Med 1999;341:1165-73.
26. Rubie H, Hartmann O, Michon J, et al. N-Myc gene amplification is a major prognostic factor in localized neuroblastoma: results of the French NBL 90 study. Neuroblastoma Study Group of the Societe Francaise d'Oncologie Pediatrique. J Clin Oncol 1997; 15:1171-82.
27. Horiuchi A, Muraji T, Tsugawa C, et al. Thoracic neuroblastoma: outcome of incomplete resection. Pediatr Surg Int 2004;20: 714-8.
28. Smith MC, Smith RJ, Bailey CM. Primary cervical neuroblastoma in infants. J Laryngol Otol 1985;99:209-14.