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NASAL CHONDROMESENCHYMAL HARMATOMA IN A NINE YEAR OLD FEMALE: A CASE REPORT

B. N. Macharia, MBChB, MMed, Senior Lecturer, Department of Pathology and Forensic Medicine, T. M. Sisenda, MBChB, MMed, FCS, Lecturer, Department of Surgery and Anaesthesiology, C. Tabitha, BSc, Moi University School of Public Health, P. O. Box 4606, Eldoret 30100, Kenya and B. C. Rono, BSN, MPH, Department of Public Health, Jomo Kenyatta University of Science and Technology, School of Public Health, P. O. Box 62000-00200, Nairobi.

Request for reprint to: Dr. B. N. Macharia, Department of Pathology and Forensic Medicine, Moi University School of Medicine, P.O. Box 4606, 30100, Eldoret, Kenya

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B. N. MACHARIA, T. M. SISENDA, C. TABITHA and B. C. RONO

SUMMARY

Hamartomas are tumorlike benign mass lesions due to excessive growth of tissues indigenous to the site of origin. Nasal chondromesenchymal hamartomas are predominantly in young infants and their occurrence in children is especially rare and only a few cases have been described in literature. Patients may present with nasal blockage leading to difficulties in breathing, rhinorrhea, epistaxis, visual disturbances, and even otitis media. Surgery is the method of treatment and the outcome is usually good when this is done early in the disease progression. We present a case of Nasal Chondromesenchymal hamartoma in a nine year old female child who presented late with a large nasal mass.

INTRODUCTION

Hamartomas are common in skin, lung, liver, chest wall, kidney, and gastrointestinal tract. In the region of the head and neck, hamartomas are uncommon but have been described in the oral cavity, nasal cavity, nasopharynx, hypopharynx, tongue, and eustachian tube (1).

The occurrence of hamartoma in the nasal cavity of infants and children is especially rare (2). By June 2014 only 31 cases had been reported (3). A hamartoma is an excessive growth of normal mature tissues native to the site it grows. However, although the cellular elements are mature they do not reproduce the normal architecture of the surrounding tissues (4).

The term nasal chondromesenchymal hamartoma was coined by McDermott *et al* in 1998, and it has since been considered a distinct clinicopathological entity. Most of the documented cases occurred in infants and it is a rare entity in children with only a few having been documented (5).

It is a benign mass lesion of the nasal cavity predominantly described in young infants and its characteristic morphology includes a proliferation of mesenchymal and chondroid or cartilaginous elements. Its pathogenesis is unknown, but it may be derived from embryologic rests (6, 7).

The largest series was recorded by McDermott *et al* and comprised 7 cases (5). Here we report such one case

CASE REPORT

Presentation: A 9 year old girl presented with difficulties in breathing and a swelling in the left nose that had been

there for one and a half years. It started off as a small mass then progressively increased. It was associated with mouth breathing and some pain.

Examination findings: The child was examined by ENT surgeon and was found to have a growth within the left nostril completely blocking it. The swelling was firm and mildly tender. The patient had a pulse rate of 97, temperature of 35.9C, and weighed 19 kgs. The child was planned for emergency examination under anesthesia (EUA), biopsy and tracheostomy. A CT- scan was also requested.

Radiology findings: The scan revealed a large antrochoanal mass.

Surgical findings: The patient was put in supine position and after administering general anaesthesia, oral endotracheal intubation was done. Oral cavity was packed. The patient was draped and the oral cavity and face cleaned. EUA showed a firm, gritty non hemorrhagic mass filling the left nostril, oropharynx, and nasal pharynx. The mass was removed piecemeal and was found to originate from the ethmoid and left maxillary sinus. The left infraorbital wall, soft palate, and nasal septal mucosa were all intact. Hemostasis was achieved. A small surgical dressing dipped in thrombin was left in the left maxillary sinus and the left nostril. The patient was successfully reversed from the general anesthesia and the mass sent for histology examination.

Pathologic findings: Grossly the tumour consisted of gray-tan

soft tissue fragments aggregating to 4cm. The cut surface was mucoid with areas of haemorrhage.

Histologically, a diagnosis of nasal chondromesenchymal hamartoma was made. It was composed of a mixture of mesenchymal tissues of spindly cells, collagen fibers, and chondroid tissue. No mitotic figures were seen.

Outcome of surgery: The patient was noted to be hyperventilating after five hours in the ward and was intubated with endotracheal tube (ETT) size 6.0mm. While a T-piece was being inserted, she developed cardiac arrest and resuscitation was done till cardiac activity was noted on ECG and by auscultation. The T-piece was inserted and the patient given 100% oxygen 7L/min but she went into cardiac arrest again. Resuscitation was unsuccessful and the patient died within one hour after she was noted to be hyperventilating.

DISCUSSION

The most common sinonasal tumors are epithelial in nature. However, nasal masses, including those of epithelial and mesenchymal origin, are infrequently encountered in young infants and children (8). The term nasal chondromesenchymal hamartoma was first coined by McDermott *et al.* in 1998 to describe a distinct clinicopathological entity composed of a proliferation of mesenchymal and cartilaginous elements (9). Before then, it has been described under various names, such as chondroid hamartoma, mesenchymoma, and nasal hamartoma (10, 11).

Exact pathogenesis of NCMH is still unknown but it is thought that it may be caused by an underlying genetic predisposition in combination with the proper stimulation by environmental factors linked to chronic inflammation or hormones.

NCMHs occur predominantly in males and infants under one year old (12, 13). However NCMH have been reported in children. In our case the patient was a female and aged 9 years. Nasal hamartomas may be predominantly composed of mesenchymal or epithelial tissues. The mesenchymal hamartomas are more common and have been named chondroid, chondromesenchymal, angiomatous, or lipomatous, depending on the preponderant tissue (14-17).

In the case we are presenting, the hamartoma was mesenchymal and mainly composed of chondroid tissue. Clinical presentation depends on the location and size of the lesion and include respiratory and feeding difficulties, rhinorrhea, epistaxis, visual disturbances, and otitis media (18). Occasionally, ophthalmologic signs such as ophthalmoplegia, proptosis, ptosis, hypotropia, and enophthalmos can result from the orbital involvement of the tumor (19). Our patient presented with nasal blockage and difficulty in breathing. This is in keeping with other reported cases. Early surgical resection is the treatment of choice and when complete resection is achieved, there is no recurrence (10, 20).

When incompletely excised reported examples of NCMH have behaved in a benign fashion (21). However it may be difficult to decide how to treat residual tumor or unresectable lesions. Local resection through functional endoscopic sinus surgery (FESS) is recommended when the tumor is localized in nasal cavity. No adjuvant therapy is necessary (22). In our case the surgical resection was done but since the child presented late with a large mass, she developed complications and died shortly after surgery.

In conclusion, although NCMH are commonly found in neonates, they can also occur in older children as we have demonstrated in this case. This is also corroborated by other case reports on the same. Early diagnosis and surgery is key for better outcome. In this case the child was referred for surgery late in the disease progression and this may have affected the outcome.

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