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(CASE REPORT)

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Nasopharyngeal fibroma presentation revealing a Burkitt Lymphoma

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Abstract

This case report details a rare presentation of Burkitt lymphoma (BL) in the naso-sinual region of a 15-year-old adolescent, which clinically and radiologically mimicked a nasopharyngeal fibroma. Initially presenting with chronic epistaxis and nasal obstruction, the patient underwent tumor resection; however, histopathological analysis revealed BL, an extremely aggressive B-cell lymphoma. Despite a rapid post-operative recurrence and the development of widespread distant metastasis, the patient achieved complete remission following a six-month intensive chemotherapy protocol. This case underscores the critical importance of considering high-grade malignancies in pediatric patients with nasopharyngeal masses, as the rapid growth potential of BL necessitates early and accurate diagnosis to prevent devastating systemic complications.

Keywords: Burkitt lymphoma; Nasopharyngeal fibroma; Epistaxis; Embolization

1. Introduction

Burkitt lymphoma (BL) is an aggressive lymphoma first described in 1958 by Denis Burkitt in Africa, in a region of endemic malaria, in an African child with a malignant mandibular tumor.[1] It is recognized as a non-Hodgkin B-cell lymphoma with high malignant potential, mainly involving extranodal lymphomas.

The annual incidence of Burkitt lymphoma in Western countries is approximately 20 to 40 times lower than in endemic regions. The non-endemic or American form of the disease differs from the original African form that its most common site of presentation is the abdomen or bone marrow.

The American form manifests in the head and neck region, but it is observed in fewer than one quarter of reported cases, and among these, it usually presents as cervical lymphadenopathy. Other sites of involvement in the head and neck region are the mandible, tonsils, jaw, and rarely the naso-sinus region.

2. Case report

A 15-year-old adolescent with no specific past medical history particularly no history of EBV, HIV, malaria, or yellow fever infection was admitted for episodes of intermittent, intense bilateral epistaxis that began 2 years ago, decompensated by iron-deficiency anemia and complicated after one and a half years by permanent right then bilateral nasal obstruction, purulent rhinorrhea, anosmia, and headaches.

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Clinical examination by nasofibroscopy revealed a mass filling both choanae and the nasopharynx, whitish, bleeding on contact, with pus draining from the middle meatus. There were no cervical or other lymphadenopathies. Ophthalmological and neurological examinations were normal.

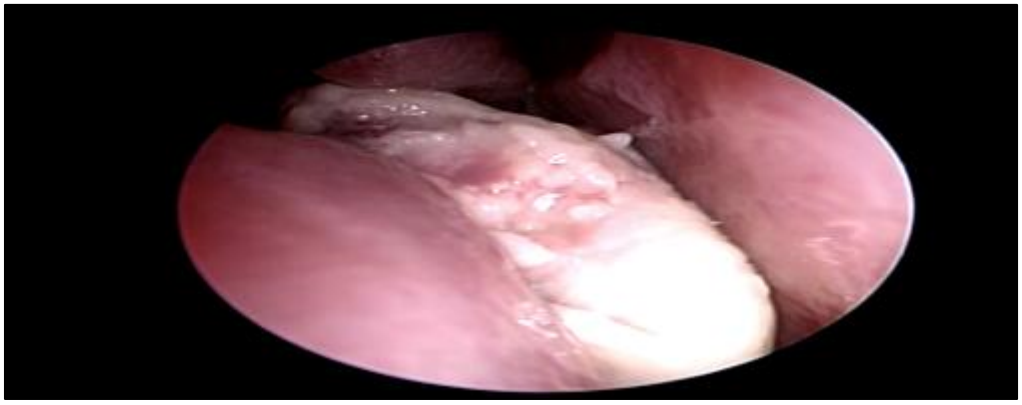

Figure 1 Endoscopic view of the tumor

Radiological workup including CT scan and MRI of the nasal sinuses was performed, revealing a sphenoid-rhinopharyngeal tumor filling both choanae, the nasopharynx, and extending to the right nasal cavity and right infratemporal fossa, with polylobulated contours, iso-signal on T1 and heterogeneous iso-signal on T2, hyperintense on diffusion-weighted imaging with ADC restriction, showing intense and heterogeneous enhancement after contrast injection.

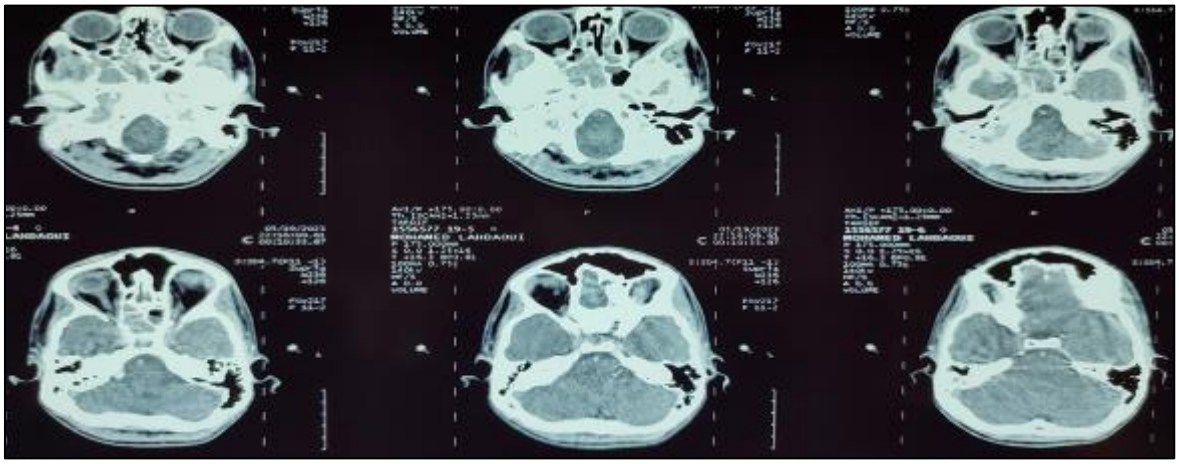

Figure 2 CT scan images of the tumor

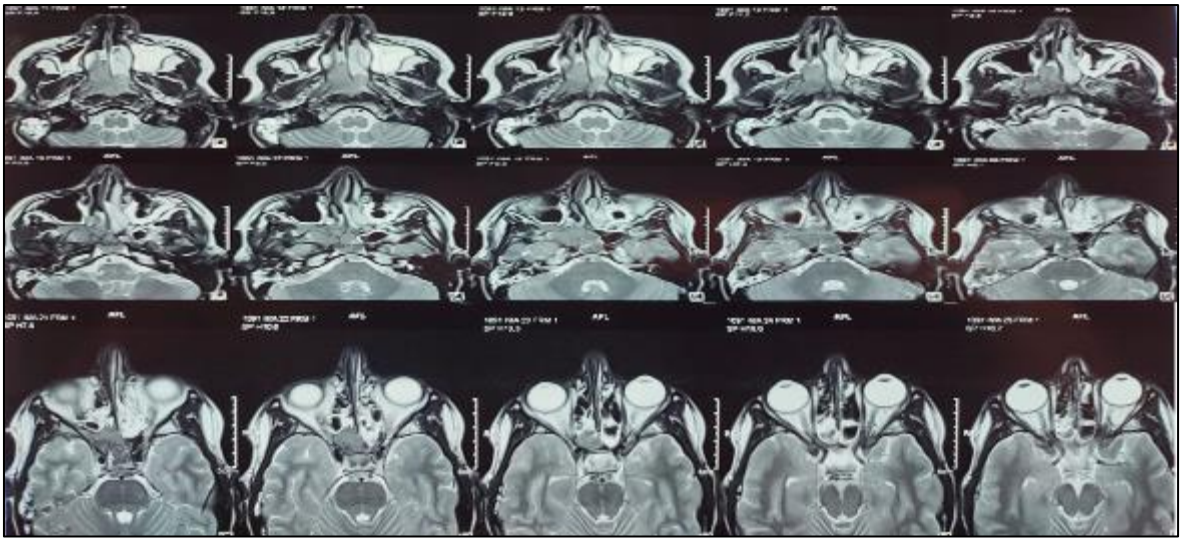

Figure 3 MRI scan images of the tumor

A staging workup was performed, revealing no loco-regional or distant metastases at that time.

Based on the clinical and paraclinical presentation described above, a diagnosis of nasopharyngeal fibroma was suggested, prompting the decision to perform angiography, which showed a typical vascular blush with a nasopharyngeal projection fed by the right internal maxillary artery.

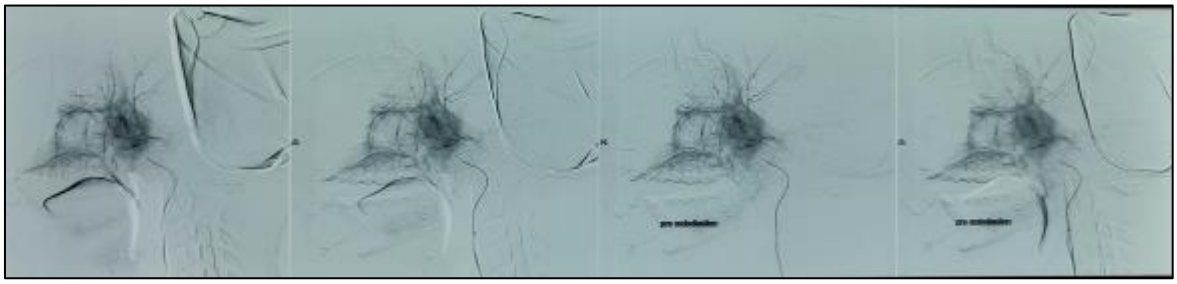

Figure 4 Angiography images showing the vascular blush

Embolization of the right internal maxillary artery was successfully performed, with near-complete disappearance of the vascular blush.

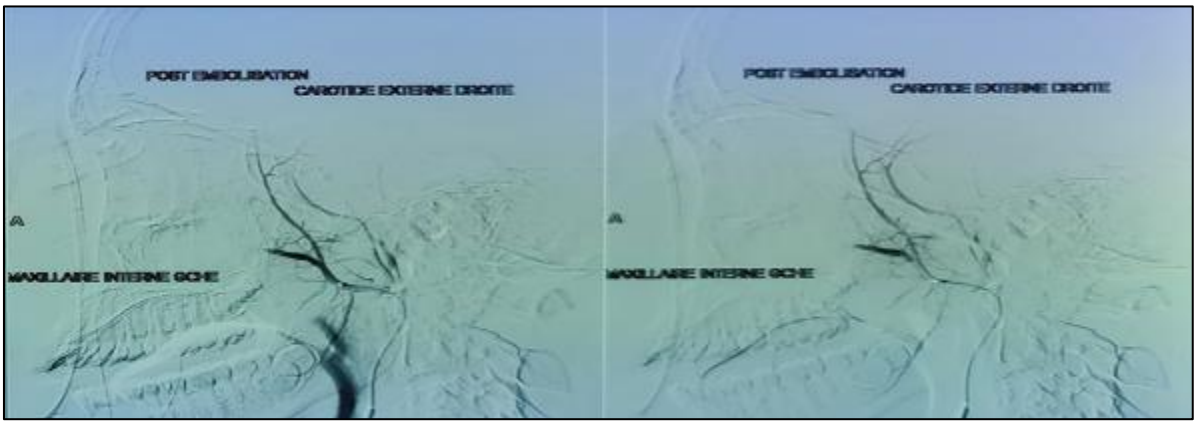

Figure 5 Disappearance of the vascular blush after embolization

A complete biological workup was performed, revealing iron-deficiency anemia with markedly elevated LDH and an inflammatory syndrome. Viral serologies were negative.

Therapeutic management consisted on a tumor resection under general anesthesia, without preoperative biopsy under local anesthesia given the hemorrhagic risk.

Resection was performed of the endonasal, sphenoidal, and pterygopalatine fossa components, with the final histopathological result: Burkitt lymphoma.

The course was marked by worsening headaches with local recurrence and the appearance of distant metastasis: meningeal, peritoneal, renal, pancreatic, pleural, paravertebral, and presacral, as well as the clinical onset of tetraplegia with cognitive impairment.

The patient was transferred to the oncology department for further management, where he received a protocol of intrathecal chemotherapy that failed after 5 sessions, followed by an intravenous chemotherapy protocol consisting of prednisone, cyclophosphamide, allopurinol, cytarabine, methotrexate, and dexamethasone, with favorable outcome and remission after 6 months.

Endoscopic and radiological follow-up at 6 months demonstrated complete disappearance of the endonasal mass and distant metastasis on CT scan.

3. Discussion

Burkitt lymphoma is a small-cell tumor with high malignant potential. Nasosinusal localization is rare, with only 14 cases reported in the past century. [1,2] Its etiopathogenesis is poorly understood, and the causal link with EBV remains unclear.[3]

BL is classified into three forms: endemic, sporadic, and immunodeficiency-associated (HIV). The sporadic and immunodeficiency types are generally found in the abdomen and bone marrow, and rarely in the head and neck region.[4] However, the endemic African type often affects the face and adjacent soft tissues and is associated with the Epstein-Barr virus.[4]

Approximately 58% of cervicofacial BL cases involve the mandible or maxilla, although very few have been reported in the nasal cavity and nasopharynx.[5] Clinically, it is suggested by the triad of nasal obstruction, purulent rhinorrhea, and headaches, with or without facial pain.[6] In our case, the patient presented with recurrent epistaxis and permanent bilateral nasal obstruction, which led to confusion with a diagnosis of nasopharyngeal fibroma.

Delayed diagnosis and treatment of BL increase the risk of distant metastasis [7] — as observed in our patient. BL has an extremely short doubling time of 24 to 48 hours.

The definitive treatment of BL is often based on the patient's age and tumor location. Various treatment options exist: radiotherapy, chemotherapy, and radioimmunotherapy.[8] The role of surgery in BL remains controversial. Surgical treatment is often necessary for decompression of the lower nasal airway, or in conditions such as optic nerve decompression if the orbit is involved, or in the context of taking a biopsy to have a confirmed diagnostic.[9]

BL responds efficiently to chemotherapy, inducing tumor regression and often leading to long-term remission. In our case, a chemotherapy protocol including prednisone, cyclophosphamide, allopurinol, cytarabine, methotrexate, and dexamethasone was administered. Currently, 5-year survival for BL in children and young adults has increased 2 to 3 times over the past three decades, from 85% to 90% with fewer than 6 months of intensive chemotherapy.[10] This is attributed to a better understanding of disease biology and advances in chemotherapeutic agents.

4. Conclusion

Burkitt lymphoma located in the naso-sinus region is an extremely rare clinical entity whose diagnosis is confirmed by histopathological and immunohistochemical examination.

Recurrent epistaxis with nasal obstruction in an adolescent, as in our case, often guides the clinician toward a diagnosis of nasopharyngeal fibroma.

Pediatricians and pediatric otorhinolaryngologists must keep in mind the diversity of clinical presentations of BL in the pediatric age group.

BL is an extremely rapidly growing tumor with a high malignant potential, necessitating early and adequate management to prevent metastasis and locoregional complications.

Compliance with ethical standards

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Disclosure of conflict of interest

The authors declare no conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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