

BURKITT'S LYMPHOMA: REPORT OF A CASE

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الدور الذي يلعبه طبيب الأسنان في تشخيص ورم بيركت اللنفاوي

يعتبر ورم بيركت اللنفاوي أحد الأورام الخبيثة التي تتميز بالنمو السريع والتشوه الشديد للوجه إضافة إلى الأجزاء الأخرى من الجسم. ويؤثر هذا الورم على كل من الأطفال والشباب.

ولقد وصف بيركت (1958م) الورم الموجود لدى أطفال أوغنديين تتراوح أعمارهم ما بين عامين وأربعة عشر عاماً. ويوجد هذا الورم في شكلين: الشكل الأفريقي والشكل الأمريكي وهناك اختلاف بين هذين الشكلين في بعض الجوانب. وما تزال أسباب هذا المرض غامضة. ومع هذا يتسبب فيروس إيبستين بار في حدوث هذا المرض منذ فترة طويلة. ويعتقد أن هذا الفيروس يعتمد في انتقاله وبقائه على العوامل المناخية الاستوائية. وقد راجع طفل سوداني عمره سبعة أعوام عيادات الطوارئ بمستشفى سنار، وكان يعاني من تورم في الجانب الأيمن من الوجه وأسنان متقلقلة وثثة متضخمة. وأجرينا تشخيصاً مبدئياً لحالة المريض حول الذروي (حول ذروة جذر السن). ولم يستجب المريض لعلاج البنسلين. وراجع العيادات مرة أخرى بعد ثلاثة أسابيع وكان يعاني من ورم منتشر بصورة متزايدة وكتلة صلبة وثثة متضخمة وأسنان متباعدة ولسان مرفوع. ولقد أدى الفحص المجهرى للنسيج إلى تشخيص ورم بيركت اللنفاوي. وقد استجاب المريض بصورة جيدة للعلاج بالأشعة.

ومن المهم أن نلاحظ أنه في المرضى الصغار الذين يعانون من مرض السن المقلقل مع وجود مرض لثوي واضح من خلال صور الأشعة، يجب أن ندرس احتمال وجود آفات خطيرة وخصوصاً إذا لم تستجب الآفات اللثوية أو حول الذروية للعلاج المناسب.

Burkitt's lymphoma is a monoclonal proliferation of B lymphoma. It is characterized by rapid growth and facial asymmetry, loosening and drifting of teeth, and enlarged gingiva. It occurs most frequently in children between the ages of 2-12 years. A 7 year-old Sudanese boy presented with a swollen face on the right side, loose teeth and enlarged gingivae. Diagnosis and treatment are presented and discussed. It is important to remember that in young patients; having a "loose tooth syndrome" with periodontal involvement confirmed by radiographic changes, one must consider the possibility of serious conditions, particularly if suspected periodontal or periapical lesions do not respond to appropriate therapy.

Introduction

In 1958 Burkitt described a tumor among children of Uganda. He noted that the tumor occurs in the age-group from 2 to 14 years with peak incidence

between 4-8 years of age. Jaw involvement is seldom in patients older than 15 years of age. The maxilla is involved more often than the mandible. However, all four quadrants of the mouth could be involved. The tumor affects males more than females. In most cases, the jaws are involved. In some patients, pancreas, kidney, abdominal lymph nodes, testes, ovaries, thyroid gland, salivary glands, breast, heart and long bones are also involved.

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O'Connor and Davies² identified the tumor as a malignant lymphoma. There are two major forms of Burkitt's lymphoma: the African and the American ones. The great majority (95%) of these tumors are seen in Africa while 5% are seen in other countries, e.g. United States, U.K., Brazil and Malaya.

The etiology of Burkitt's lymphoma is obscure. However, Epstein-Barr virus has long been associated with Burkitt's lymphoma. It appears to thrive in tropical climates where optimal conditions exist for its survival. A further possible cause is chromosomal abnormalities. Yih *et al*³ stated "despite the difference in clinical epidemiologic and serologic features of these forms, both tumors do share another feature in common. They frequently express the highly specific cytogenetic t(8;14) marker. This event is the result of a reciprocal translocation between chromosomes 8 and 14, and the site of breakage is identical in most cases of Burkitt's lymphoma, whether African or American in origin".

De The⁴ suggested that perinatal or neonatal Epstein-Barr virus infection may predispose a child to development of Burkitt's lymphoma. A further possibility is an altered immune response caused by chronic infection which may promote the oncological effect of the tumor. It has also been postulated that chronic malarial infection may play a role in the development of the lymphoma.⁵

The first sign of Burkitt's lymphoma is loosening of the deciduous molars which drift and become displaced. Eventually the teeth are exfoliated because of the severe bone loss. Other clinical signs include asymmetry of the face, paresthesia and pain. However, radiographically, early signs include an area of osteolysis in connection with the germs of the developing teeth (crypts) on the other hand later sign shows an extensive osteolysis and floating teeth phenomena. Such changes can be detected by orthopantomogram (OPC) and periapical views.

The aim of this paper was to review the important role of the dentist in the diagnosis and treatment of Burkitt's lymphoma.

Case Report

A 7-year-old Sudanese boy presented to the outpatient clinic at Sennar Hospital in Sudan. His chief complaint was swelling of the right side of the

face. On examination, there was a loose tooth in the molar area. The gingiva was inflamed and enlarged. A provisional diagnosis of periapical infection was made. The patient was treated with penicillin and appointed to return in one week. Unfortunately he did not keep his appointment and six weeks later he returned with a markedly increased, diffuse, firm swelling of the right face extending to the inferior border of the orbit [Figs. 1 and 2]. Many of the teeth had become mobile and drifted. One of the molars was exfoliated out of its socket. The tongue was elevated. The patient was unable to close his mouth, chew or swallow. His general appearance was poor and appeared anaemic and was admitted to the hospital. The admitting haemoglobin was 2.8 gm/ml. A nasogastric tube for feeding was inserted. The patient received several units of blood. A differential diagnosis of ameloblastoma, Burkitt's lymphoma, fibrosarcoma or Ewing sarcoma was established.



Fig. 1. Patient with massive swelling of the mandible.



Fig. 2. Closer view of the growth.



Fig. 3. Histologic examination showing the "starry sky" appearance.

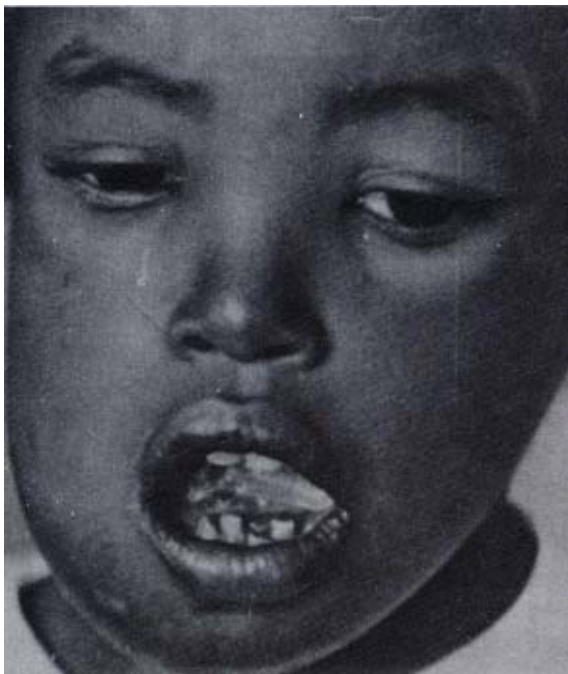


Fig. 4. Patient after one week of treatment.

The patient was referred to Khartoum Teaching Hospital. A biopsy was taken and the histopathological examination revealed the classic starry sky pattern which is pathognomonic of Burkitt's lymphoma [Fig. 3].

The patient was treated with radiotherapy. After one week a remarkable decrease in the swelling was observed [Fig. 4]. He was discharged and given an appointment to return for follow-up which he failed. Few months later, he died because of secondary metastasis.

Discussion

In its early stages of development Burkitt's lymphoma can be very difficult to diagnose. Patients generally present with complaints and symptoms that mimic common dental problems i.e. advanced caries with pulpal degeneration leading to a chronic periapical abscess that suddenly becomes acute. The dental practitioner must evaluate the extent of caries, the amount of periapical and periodontal bone loss and consider other pathologies particularly if caries is absent. If after the initial assessment there is no change in the lesion in a 2-week period; a biopsy must be performed. It would be prudent to have a complete blood analysis including haemoglobin, hematocrit, differential count and blood chemistry. Additionally, a biopsy of the abnormal gingival tissue would be most important. When extraction is indicated in young children with no history of periapical or periodontal infection, a biopsy should be taken from the extraction site, kept in 10% formalin and sent to pathology. It is critically important to establish a diagnosis as soon as possible since the prognosis is favorable with an early diagnosis.

Burkitt's lymphoma is very responsive to chemotherapy as Burkitt described.⁶ The chemotherapy protocol can include cyclophosphamide (Cytoxan), vincristine (Oncovin), methotrexate sodium, and prednisone. Radiation therapy appears to be a very good alternative. The role of radiotherapy is unknown, but less likely to be effective in multicentric lesions. Our patient responded very well to radiation therapy and most of the tumor disappeared [Fig. 4].

Early diagnosis is most important in the prognosis of Burkitt's lymphoma, otherwise metastasis may

develop very rapidly in other organs (spinal cord, liver, bone marrow etc..) and the prognosis becomes unfavorable with high mortality rate.

A team effort by the clinician, oncologist, hematologist, and radiotherapist is very important in the treatment and foliow-up since relapse have been reported in some patients.

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