Case report/Kazuistyka

Unilateral non-Hodgkin's lymphoma of the frontal sinus presenting as Pott's puffy tumour

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Introduction

In the Western world, malignant lymphoma of the paranasal sinuses and nasal cavity is rare with an incidence ranging from 0.17% to 1.63% [1–3] and only account for 0.44% of extranodal lymphomas [1]. These sinonasal lymphomas arise from lymphoid tissues found in the sinus submucosa or bone marrow [4].

It usually presents as an isolated disease of the sinuses [5] which is normally non-specific initially [3, 6], or locally advanced lesions involving adjacent structures [7], and rarely present as concurrent generalised lymphoma [5]. Therefore, it can mimic those of infectious, non-lymphomatous neoplastic and granulomatous process in their initial presentation [8], thus delaying its diagnosis [6].

In this article we will present a case of unilateral frontal sinus B-cell lymphoma which was treated as Pott's puffy tumour. Key features to differentiate the two conditions are discussed. A 61-year-old woman presented with 4 weeks history of a gradually enlarging right-sided forehead swelling, which was smooth and firm with no tenderness, fluctuation or overlying skin changes.

Initial computed tomography (CT) of the paranasal sinuses revealed a mildly opacified right frontal sinus with an overlying soft tissue swelling. A repeat CT scan one week later showed evidence of osteomyelitis of the medial wall of the right frontal bone and orbit, suggesting diagnosis of Pott's puffy tumour. A third CT scan was performed as the swelling continued to enlarge despite antibiotics, which demonstrated a collection over the frontal sinus. No pus was found from a frontal sinus trephine. Histopathologic analysis of the frontal sinus biopsy showed diffuse large B cell lymphoma. The patient was treated successfully with 6 cycles of chemotherapy. In conclusion, it is not easy to differentiate Pott’s puffy tumour from frontal sinus lymphoma at their initial presentation. With clearer understanding of both conditions, we can raise the index of suspicion among clinicians of the possibility of frontal sinus lymphoma even when evidence of osteomyelitis is present, therefore prompting earlier tissue biopsies for confirmation of diagnosis.

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tumour initially, with the intention to raise awareness of sinonasal lymphoma and discuss the key differences between the two conditions to help establish the correct diagnosis earlier.

Case report

A 61-year-old woman presented with 4 weeks history of a gradually increasing right-sided forehead swelling which was only mildly painful. There were no associated symptoms of nasal congestion, pyrexia or visual disturbances. She had a history of recurrent sinusitis about twice a year, which normally settled with oral antibiotics. Her past medical history also included hypertension, previous tonsillectomy and Fallopian tube ligation. She had never smoked.

On physical examination, the patient was found to have a 4 cm × 4 cm swelling over the right frontal sinus, which was smooth and firm with no tenderness, fluctuation or overlying skin changes. Flexible nasendoscopy was unremarkable.

Computed tomography (CT) of the paranasal sinuses revealed a subcutaneous soft tissue swelling overlying the right frontal sinus with no evidence of bony destruction or intra-orbital mass lesion. The frontal sinus was mildly opacified. She was discharged with oral Clarithromycin and topical nasal decongestants with a working diagnosis of frontal sinusitis with superficial cellulitis.

She presented to the ENT clinic again one week later where the swelling had increased in size to 5 cm × 5 cm. Otherwise, there were no changes to its characteristics on physical examination. She was admitted to the hospital for treatment with intravenous Co-Amoxiclav and topical nasal decongestants. Endoscopic drainage of right frontal sinus was attempted, but no pus was identified intra-operatively.

As her headache worsened with associated vomiting a day later despite the surgical intervention, CT scan of the brain and the paranasal sinuses was repeated. In addition to the persistent right frontal subcutaneous soft tissue swelling, the scan showed opacification within the right frontal, anterior ethmoidal and maxillary sinuses. Bony rarefaction of the medial wall of right frontal bone and orbit suggestive of osteomyelitis was also reported (Fig. 1). The diagnosis of Pott’s puffy tumour was suggested.

However, the frontal swelling continued to increase in size despite antibiotics and a third CT scan was performed to demonstrate a collection in the frontal sinus (Fig. 2). The patient had a right frontal sinus trephine on the same day, but again no collection was found. Frontal sinus biopsies were taken. Fluid culture from the frontal sinus showed light growth of Serratia Marcescens. Therefore, based on microbiological guidance, intravenous Piperacillin/Tazobactem therapy was commenced. Nevertheless, her symptoms continued to deteriorate and the patient developed ptosis from the mass.

Ultimately, histopathologic analysis of the mass showed diffuse large B cell lymphoma. Subsequent staging CT showed localised disease only.

Throughout her admission, all the haematological and biochemical investigations had been normal.

She is currently undergoing 6 cycles of chemotherapy with Cyclophosphamide, Doxorubicin, Vincristine and Prednisolone, and intrathecal Methotrexate for central nervous system prophylaxis.

Discussion

The initial diagnosis for our patient was Pott’s puffy tumour, a condition first described by Sir Pervical Pott in 1775 as a circumscribed, puffy swelling of the forehead or scalp associated with osteomyelitis of the frontal bone and intracranial infection [9]. This condition usually presents as a unilateral mass or swelling in the forehead associated with fever and frontal headache [9–12]. The swelling is usually described as smooth, doughy [10] or fluctuant [12] with surrounding cellulitis and peri orbital oedema. It is caused by a subperiostal abscess secondary to the underlying frontal bone osteomyelitis [10]. Pott’s puffy tumour usually occurs as a complication of frontal sinusitis [10, 11, 13, 14], trauma [14] or late complication of frontal sinus reconstruction [14]. Although it affects mostly teenagers, there have been cases reported in patients aged from 7 to 83-year-old [12]. CT scan of the sinus usually shows subperiosteal abscess and pansinusitis [10–12] with frontal bone osteomyelitis [12] or erosion [11]. Common causative organism includes Streptococcus, Staphylococcus and anaerobes [9].
Pott’s puffy tumour may lead to further complications of fronto-cutaneous fistula and intracranial extension of the infection [13]. However, Pott’s puffy tumour has become a rare occurrence due to extensive usage of antibiotics, early lavage of sinuses after lack of response to medical therapy and usage of imaging to aid early diagnosis [13].

In our case, the patient fulfilled majority of the typical features of Pott’s puffy tumour. She had a history of recurrent sinusitis and presented with a unilateral lump over the frontal sinus. Her CT scan showed pansinusitis with evidence of osteomyelitis. However, there were no history or signs of recent or current sinusitis and she denied any trauma to the forehead. On clinical examination, the lump was hard, rather than “puffy”, fluctuant or doughy. The swelling continued to grow in size despite treatment with intravenous antibiotics. The culture taken from frontal sinus lavage showed an atypical organism, Serratia Marcescans, and there were no pus drained on both trephine procedures. Her CT scan has been reported as “soft tissue mass” in the frontal sinus.

In comparison, frontal sinus lymphoma usually presents as an isolated disease of the sinuses [5] or locally advanced lesions involving adjacent structures [7], and rarely present as concurrent generalised lymphoma [5]. As shown in our case, it can mimic the features of infectious, granulomatous or non-lymphomatous neoplastic pathologies in their initial presentation [8], thus delaying the diagnosis [6].

Sinonasal lymphoma is more common in Asian population [15] with an incidence of 6.7% of all lymphomas where it is the second most common extranodal lymphoma after gastrointestinal lymphomas [16]. The incidence is also higher in South American countries [15]. Histopathological difference in the sinonasal lymphomas exist between different ethnic groups. In the Western population, B-cell lymphoma is more common [15] and mostly affects the maxillary antrum [15], nasal cavity and the ethmoid sinuses [7]. On the other hand, T-cell phenotype is commoner among Asian population [3, 16] where it predominantly affects the nasal cavity [2, 16].

Although it is common for patients to present with ophthalmological features such as proptosis, eye pain and diplopia in the early stage of the disease due to the close proximity of the paranasal sinuses to the orbit [17], symptoms of frontal disease is normally non-specific initially [3, 6]. External presentations of the disease like cellulitis, bony destruction and oedema are typically late signs [6]. Wilder et al. [8] divided the symptoms into three categories, which include local symptoms (rhinorrhea, epistaxis, obstruction), symptoms of adjacent structures invasion (ear fullness and facial pain) and systemic symptoms (weight loss, nocturnal sweating and fever).

Early diagnosis allows effective treatment [8]. However, it is difficult to evaluate the frontal sinus intranasally or endoscopically due to the anatomic constraints [6]. Diagnosis therefore rely on clinician’s awareness and high index of suspicion when patients present with these features on physical examination, with the help of computed tomography (CT) or magnetic resonance imaging (MRI) of the sinus [6] and surgical biopsy [3]. When performing a tissue biopsy, it is important to remember that lymphoma is present in the submucosal tissues and thus there may not be an obvious lesion to biopsy on endoscopic inspection. The specimen sent should be fresh in order to follow the surgical pathologic lymphoma protocols rather than in formaldehyde [16].

**Conclusion**

It is not easy to differentiate Pott’s puffy tumour from frontal sinus lymphoma at their initial presentation. With clearer understanding of both conditions, we can raise the index of suspicion among clinicians of the possibility of frontal sinus lymphoma even when evidence of osteomyelitis suggesting Pott’s puffy tumour is present, therefore prompting earlier tissue biopsies for confirmation of diagnosis.

**Authors’ contribution/Wkład autorów**

EW – study design, data interpretation, acceptance of final manuscript version, literature search. W-YY – data interpretation, acceptance of final manuscript version. DL – acceptance of final manuscript version.

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Ethics/Etyka

The work described in this article have been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; EU Directive 2010/63/EU for animal experiments; Uniform Requirements for manuscripts submitted to Biomedical journals.

The own research were conducted according to the Good Clinical Practice guidelines and accepted by local Bioethics Committee, all patients agreed in writing to participation and these researches.

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References/Piśmiennictwo


