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Paranasal sinusitis complicated by Pott's puffy tumour

Zapalenie zatok przynosowych powikłane guzem Potta

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Abstract

Pott's puffy tumour is defined as inflammation of the frontal bone marrow with an accompanying subperiosteal abscess manifested by swelling of the frontal region and is most often a complication of paranasal sinusitis or trauma. Early diagnosis and implementation of proper treatment are important due to the frequent co-occurrence of intracranial complications, such as epidural empyema, cerebral abscesses or meningitis. Due to the low prevalence of Pott's puffy tumour, the knowledge on this severe complication relies on descriptions or series of cases, especially among children. We present a case of an 8-year-old girl who was admitted to the hospital due to headache and swelling in the frontal area. We also present a summary of the available publications on Pott's puffy tumour. Diagnostic procedures revealed massive inflammatory changes in the paranasal sinuses, with the destruction of the frontal sinus walls, and showed a connection between the inflammatory lesions and the anterior cranial fossa. The girl was qualified for urgent neurosurgical and laryngological surgery.

Keywords: frontal swelling, complication of rhinosinusitis, Pott's puffy tumour

Streszczenie

Zapalenie szpiku kostnego kości czołowej z towarzyszącym ropniem podokostnowym manifestującym się obrzękiem w okolicy czołowej definiowane jest jako guz Potta i najczęściej stanowi powikłanie zapalenia zatok przynosowych lub urazu. Wczesne ustalenie rozpoznania i wdrożenie właściwego leczenia jest istotne ze względu na częste współwystępowanie powikłań wewnątrzczaszkowych, takich jak ropniak nadtwardówkowy, ropnie mózgu czy zapalenie opon mózgowo-rdzeniowych. Ze względu na niewielką częstość występowania guza Potta zakres wiedzy dotyczącej tego ciężkiego powikłania stanowią opisy przypadków lub serie przypadków, zwłaszcza wśród dzieci. Prezentujemy opis przypadku 8-letniej dziewczynki, która zgłosiła się do szpitala z powodu bólu głowy i obrzęku w okolicy czołowej, oraz podsumowanie dostępnych wiadomości dotyczących guza Potta. Postępowanie diagnostyczne u dziewczynki wykazało masywne zmiany zapalne obejmujące zatoki przynosowe z destrukcją ścian zatok czołowych oraz komunikację zmian zapalnych z przednim dołem czaszki. Dziewczynka została zakwalifikowana do pilnej interwencji neurochirurgicznej i laryngologicznej.

Słowa kluczowe: obrzęk w okolicy czołowej, powikłanie zapalenia zatok przynosowych, guz Potta

INTRODUCTION

Although severe complications of sinusitis, such as brain abscess, osteomyelitis, meningitis and cerebral venous sinus thrombosis, are increasingly rarely encountered in the era of antibiotic therapy, they may contribute to increased mortality among children if undiagnosed. Pott's puffy tumour (PPT) is defined as a subperiosteal abscess accompanied by bone marrow inflammation, which develops as a complication of frontal sinusitis. It is one of the most severe complications of sinusitis.

We present a case of an 8-year-old girl with Pott's puffy tumour as well as a summary of the available knowledge based on a literature review.

CASE REPORT

A previously healthy 8-year-old girl was admitted to the Department of Paediatrics in the "Bielański" Hospital in Warsaw due to headache and oedema of the upper eyelids and frontal region.

The patient reported frontal headache for 2 weeks. There was poor response to analgesics (paracetamol and ibuprofen); the pain woke the girl from sleep twice. Increasing pain in the frontal and eyelid region was observed for 3 days. Two days before admission, the patient was consulted by a pediatrician, an allergic reaction was diagnosed, ibuprofen and desloratadine were prescribed – without improvement. The girl complained of nasal obstruction, drainage of discharge down the back of the throat and impaired taste sensation lasting for one day. No fever, vomiting or nausea were reported. Head trauma and insect bite were also excluded. She dived in the Adriatic Sea 4 weeks prior to admission.

The girl was in good overall condition except for a painful, soft swelling with a size of 4×5 cm, located in the frontal area and accompanied by bilateral, asymmetric oedema of the eyelids, convergent squint, reddened conjunctiva, nasal obstruction and congested palatine-pharyngeal arches. Neurological examination showed no abnormalities except for slightly positive Romberg's test.

Laboratory tests revealed slightly increased C-reactive protein (CRP) levels (11.42 mg/L vs. <5 mg/L) and white blood cells (WBC) count (16.8 thousand/ μ L). Other than that, no other abnormalities were found. A suspicion of paranasal sinusitis was raised.

Computed tomography (CT) of the sinuses revealed massive inflammatory lesions filling the entire right maxillary sinus, the right anterior ethmoidal sinuses with thinning of osseous laminae as well as the entire frontal sinus and segmental destruction of the exterior and interior wall of the frontal sinus. The inflammatory lesions were in direct contact with the anterior cranial fossa. Furthermore, thickened soft tissue in the frontal region adjacent to the frontal sinus, with a thickness of 7 mm as well as inflammatory lesions involving the right middle nasal concha with a narrowed right nasal passage were shown (Figs. 1–3).

Paranasal sinusitis complicated with Pott's puffy tumour was diagnosed based on the clinical picture and laryngological consultation. The patient was transferred to a specialist department due to the need for urgent neurosurgical intervention and laryngological treatment.

POTT'S PUFFY TUMOUR IN THE PAEDIATRIC POPULATION

Pott's puffy tumour is described as inflammation of the frontal bone marrow associated with the collection of purulent

Fig. 1. CT (bone window), frontal view: massive inflammatory lesions involving the right maxillary sinus (a) and the anterior ethmoidal sinuses with thinning of osseous laminae (b)

Fig. 2. CT (bone window), cross-sectional view: destruction of the anterior and posterior wall of the frontal sinus (a) and soft tissue oedema in the frontal region (b)

Fig. 3. CT (bone window), sagittal view: inflammatory lesions filling the frontal sinus (a), destruction of the posterior wall of the frontal sinus (b) and soft tissue oedema in the frontal region (c)

content in the subperiosteal space, clinically manifested as a puffy swelling and oedema located in the frontal area⁽¹⁾. It is one of the most severe consequences of untreated or inadequately treated sinusitis, which may lead to severe intracranial complications⁽²⁾.

The vast majority of publications on Pott's puffy tumour are case reports or case series, with children being the largest group of patients⁽³⁾. A total of 115 cases, including 74 children and 41 adult patients, had been described by 2017 (data from the PubMed database – 1960–2017). Below, we present a detailed characteristic of the disease. The following information summarises the available knowledge and may be of help in this rare diagnosis.

Aetiology

Pott's puffy tumour usually develops as a complication of chronic or acute frontal sinusitis^(1–20). In most cases, the diagnostic process reveals inflammation of more than one sinus, with maxillary and ethmoidal sinuses being a common location. Up to 75% of patients with Pott's puffy tumour have a medical history of acute or chronic sinusitis⁽⁴⁾. It is estimated that 15% of paediatric patients develop the tumour following head trauma with no connection with sinusitis⁽⁵⁾. Other causes described in the worldwide literature include craniotomy, mastoiditis, insect bite, otitis media, acupuncture, periodontitis, hair transplantation, cocaine/methamphetamine abuse, and a retention cyst of the frontal sinus^(5–7).

Epidemiology

Although Pott's puffy tumour occurs in all age groups, it mostly affects adolescents, with a male predominance (3:1)^(8,9).

The mean age at the diagnosis is 11.6–11.8 years, which is assumed to be due to the higher frequency of upper respiratory tract infections leading to sinusitis and the specific structure of frontal sinus at this stage of development^(1,5). The youngest case described in the literature was a 7-week-old child born at 25 weeks gestation, who developed Pott's puffy tumour after methicillin-resistant *Staphylococcus aureus* sepsis; however, apart from casuistic reports, children over 5 years of age represent the group of patients with Pott's puffy tumour⁽⁹⁾.

Pathogenesis

The frontal sinus originates from the anterior ethmoidal cells and develops between the internal and external laminae of the frontal bone during the process of pneumatization. Frontal sinus pneumatization begins around the age of 2 years and lasts until 12–15 years of age. The avascular diploic veins also reach the target size at this age^(5,9,10). The frontal sinus becomes radiographically visible between 6 and 8 years of life^(5,11).

Pott's puffy tumour usually develops as a complication of spreading frontal sinusitis leading to frontal bone marrow inflammation^(1–20). The direct anterior spread of frontal sinus infection causes destruction of the frontal bone cortex and subperiosteal collection of purulent content, which results in a distinct swelling in the frontal region. If the infection spreads towards the posterior wall of the frontal sinus, complications such as meningitis, epidural and subdural abscesses may develop^(1,5,9). If the bottom wall of the sinus is destroyed, complications within the eye and orbit are observed^(3,12).

Hematogenous spread is another potential mechanism of the spread of infection. The sinus mucosa is drained by the avascular diploic veins, which form anastomoses with the subdural venous plexus and the periosteal veins. Infected material may travel through the veins, destroying bone structure and leading to bone marrow inflammation with subdural or intracerebral abscess as well as encephalitis and venous sinus thrombosis^(5,8,9).

Clinical manifestation

Pott's puffy tumour is typically manifested as a specific set of subacute symptoms, with a typical swelling over the forehead. A characteristic picture seen in the course of Pott's puffy tumour is a well-delineated soft and painless tumour located on the forehead or top of the head, covering the area of osteomyelitis of the frontal bone^(1–20). Other most common symptoms are less specific and include headache (88% of patients) and fever, nasal obstruction, periorbital oedema and fatigue^(9–11,13–15). Due to the non-specificity of the initial symptoms, the use of antibiotic therapy may mask or delay the symptoms of intracranial spread of infection⁽¹⁶⁾. Symptoms indicating possible intracranial complications are seen in 30–40% of patients and include

apathy, convulsions, impaired consciousness, blurred vision, aphasia, symptoms of cranial nerve palsy, nausea and vomiting^(5,9). Orbital involvement is observed in 30% of patients and may manifest as eyelid oedema, inflammation of the connective tissue and eye movement disorders⁽⁵⁾.

Differential diagnosis

The following conditions are included in the differential diagnosis of frontal swelling in a child: sinusitis, angioneurotic edema, insect bite, Pott's puffy tumour, orbital soft tissue inflammation, multiple furuncle, atheroma, hematoma, subgaleal haemorrhage, lipoma, liposarcoma, pseudoaneurysm of superficial temporal artery, nephrotic syndrome (for general characteristics, see Tab. 1)^(14,17,18). Thorough physical examination and medical history are essential for a correct preliminary diagnosis and initiation of the diagnostic process.

Additional diagnostic tests

Laboratory abnormalities are closely related to disease progression and may be even absent at an early stage. The most common abnormalities include increased white blood cell

count and CRP levels. However, these are non-specific symptoms. Other biochemical parameters usually remain normal^(5,15). Collection and analysis of cerebrospinal fluid is of no use in the diagnosis and treatment monitoring due to the absence of specific abnormalities. Suspected meningitis is the only indication for cerebrospinal fluid collection [diagnostic imaging (CT or magnetic resonance imaging – MRI) should be performed prior to the procedure due to possible presence of other complications that may increase the intracranial pressure]⁽³⁾.

CT is a standard imaging technique to confirm the diagnosis⁽⁵⁾. In most cases, a CT scan shows an inflammatory process involving several sinuses. MRI is recommended only for suspected intracranial complications due to lower availability and the time needed for both the procedure itself and radiological assessment. However, MRI is more precise and provides better imaging of the progression of potential complications^(12,14). The sensitivity of MRI vs. CT for detecting intracranial abscess is 100% vs. 92%, respectively⁽¹⁴⁾. Although ultrasonography allows for the detection of subperiosteal infiltration and frontal bone destruction, it is of no use for assessing intracranial complications. Therefore, ultrasonography may be used only for the preliminary diagnosis after a suspicion of Pott's puffy tumour is raised⁽⁵⁾.

Diagnosis	Definition	Distinguishing symptoms
Sinusitis	Inflammation of the mucosa of one of the paranasal sinuses with symptoms of rhinitis	Diffuse swelling, purulent discharge from the nasal passages associated with facial pain and tenderness in the sinus projection
Angioedema	Subcutaneous tissue edema due to dilation and increase in blood vessel permeability	Develops within a few minutes to several hours, asymmetric and well-delineated, most often in the region of the eyelids
Insect bite	Reaction after an insect bite associated with immunological or non-immunological mechanisms	Rapidly growing oedema accompanied by erythema at the bite site, pain, burning and bursting sensation. Symptoms usually resolve within 24–48 hours
Pott's puffy tumour	Subacute subperiosteal abscess, most often a complication of sinusitis or frontal trauma	Well-delineated, soft, localised swelling accompanied by headache, mucus secretions in the nasal passages, fever and neurological manifestations
Orbital soft tissue inflammation	Skin and subcutaneous tissue bacterial infection, usually caused by upper respiratory infection, trauma or insect bite	Severe erythema, oedema and increased warmth of the eyelids, possibly accompanied by exophthalmos, blurred vision and limited eye mobility
Multiple furuncle	Suppurative perifollicular inflammation with the formation of a necrotic plug	High tenderness of the lesion, erythema, a swollen area on the skin with a pustule located on its top, under which dead tissue is accumulated
Atheroma	A retention cyst formed within the hair follicle and sebaceous gland	Slow, long-term growth, painless nodule with smooth surface, movable. It may develop bacterial superinfection and inflammation of the adjacent tissues
Haematoma	Massive extravasation of blood into tissues	Sudden onset oedema and subcutaneous blood extravasation due to injury or wounding in the involved area
Subgaleal haemorrhage	Bleeding located between the scalp and the skull, which may indicate vascular malformation or blood coagulation disorders	Unstable marshy mass, which slowly increases in the skull projection and extends beyond cranial sutures
Lipoma	A benign tumour developing from the adipose tissue	A very slowly growing soft tumour, more compact than the surrounding tissue, movable, painless, the skin covering the lesion is unchanged
Liposarcoma	A malignant tumour originating from adipocytes	Rapid growth, non-movable, painless, soft, lobular
Pseudoaneurysm of superficial temporal artery	Develops as a result of rupture of the arterial wall surrounded closely by muscles, fascia and connective tissue. Usually due to trauma	Palpable pulsation of the lesion consistent with the heart rhythm
Nephrotic syndrome	A clinical condition characterised by hypoproteinaemia leading to oedema	Symmetrical, soft swelling, its location depends on the body position – in the morning frequent swelling of the face, especially within the eyelids

Bacteriology

In each case of suspected Pott's puffy tumour, appropriate specimens should be ensured by collecting discharge from the nasal cavity and performing a puncture of the tumour cavity or aspiration of the pus from the sinus for microbiological evaluation^(11,19). It is believed that *Streptococcus*, *Staphylococcus*, *Enterococcus* and anaerobic bacteria such as *Fusobacterium* and *Bacteroides* are the main pathogens responsible for infection complicated by Pott's puffy tumour⁽¹⁴⁾. Gram-negative bacteria (*Haemophilus influenzae*) were the causative factor in several case reports^(9,20). Compared to other respiratory infections, most of complications are caused by anaerobic bacteria due to the low oxygen levels in the frontal sinus, which results from impaired patency of the ostium of the frontal sinus. Mixed bacterial flora accounts for the 2/3 of cases of positive bacteriological cultures⁽⁵⁾.

Intracranial complications

Patients with Pott's puffy tumour are a high-risk group for intracranial complications, which include the collection of purulent content in the epidural or subdural space or cerebral tissue as well as meningitis, encephalitis, venous sinus thrombosis or stroke with the formation of ischemic necrotic area^(3-5,7). They are significantly more common among children compared to adults^(8,20). According to reports, 80–85% of paediatric patients already had intracranial complications, epidural abscess in most cases, at the time of diagnosis of Pott's puffy tumour^(2,5,10,13). It is necessary to be aware of the risk of complications as the symptoms may be initially discreet or absent, especially at an early stage, or masked by previously included treatment⁽³⁾.

Treatment

A two-stage treatment is used. The first stage involves broad-spectrum antibiotic therapy, while the second stage involves neurosurgical intervention. Due to the limited available data, there are no recommended methods of pharmacological or neurosurgical treatment.

Initial diagnosis and blood sampling for microbiological tests should be followed by empirical intravenous antibiotic therapy, considering the good penetration across the blood–brain barrier and effective action on anaerobic bacteria⁽⁹⁾. Generally, third generation cephalosporins, metronidazole and vancomycin are recommended^(3,12). Pharmacological treatment should be modified, i.e. targeted antibiotic therapy should be included, after obtaining the results of microbiological analysis. Parenteral treatment should last not less than 6–8 weeks^(10,12-14). The main aim of neurosurgical treatment is to evacuate purulent content and remove bone fragments with focal bone marrow inflammation. Therapeutic management will depend on individual cases. Sinus irrigation, frontal craniotomy with evacuation of purulent content and partial excision of frontal bone are the most commonly chosen methods.

Although only one neurosurgical intervention is usually needed, another, more radical surgery may be necessary in some cases⁽¹⁴⁾.

Prognosis

Although the vast majority of patients recover without any neurological disorders, early and correct diagnosis is of key importance. Possible long-term complications include convulsions, aphasia and paralysis of different muscle groups. Their frequency is closely related to disease severity. Due to the wide availability of pharmacotherapy in the form of targeted antibiotic therapy, the mortality is low, i.e. 17% of the described cases⁽⁵⁾.

DISCUSSION

The presented case shows that early and correct diagnosis is of key importance in the case of suspected complication of paranasal sinusitis in a child.

The duration of the patient's symptoms was relatively short (2 weeks of headache in the frontal region, 3 days of frontal swelling), with typical symptoms of paranasal sinusitis, such as nasal obstruction and discharge running down the back of the throat, which persisted for one day. The girl was not feverish. These scarce clinical manifestations were the reason for the delayed medical appointment and an initially incorrect diagnosis (allergic reaction). This delay contributed to the development of frontal bone marrow inflammation and full-blown Pott's puffy tumour. This was a well-delimited, soft tumour located in the frontal region and accompanied by a periorbital oedema. It covered the area of frontal osteomyelitis. The patient complained of headache, nasal obstruction and fatigue.

Medical history revealed impaired taste sensation. Physical examination showed a positive Romberg's test, which could have suggested intracranial complications. The girl was in the age group typical for the diagnosis of Pott's puffy tumour. Chronic, undiagnosed oligosymptomatic sinusitis, which increased after swimming in the Adriatic Sea, was the probable cause.

The diagnostic process (laboratory tests and CT) revealed:

- minor increase in inflammatory markers;
- characteristic, massive lesions in CT images.

No other abnormalities were found.

After establishing the diagnosis, a decision was made to refer the patient to a specialist department for simultaneous conservative and surgical treatment. Early diagnosis and urgent targeted treatment are necessary to achieve full therapeutic success and reduce the risk of distant consequences or complications.

CONCLUSIONS

Pott's puffy tumour is a rare and, at the same time, serious complication of sinusitis. Despite the widely used antibiotic therapy and proper treatment regimens in sinusitis, there are

an increasing number of cases of Pott's puffy tumour reported in the literature. This may be explained by advances in imaging technology, obviously facilitating the diagnosis, as well as increased alertness of paediatricians who are the first to deal with these patients. The diagnosis of Pott's puffy tumour is important due to the high risk of complications and the need for urgent multidisciplinary treatment involving paediatricians, laryngologists, neurosurgeons and ophthalmologists.

Conflict of interest

Authors do not report any financial or personal connections with other persons or organisations, which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

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