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NEUROLOGICAL SYMPTOMS DERIVED FROM ISOLATED SPHENOID SINUSITIS IN CHILDREN: A REPORT OF 3 PATIENTS

MARIA FARINI, STEFANIA TOTSKA, CATHERINE MONTI*

Inflammation of the sphenoid sinus is rare, and usually occurs in conjunction with other paranasal sinus infections. Isolated sphenoiditis is even rarer and is reported almost uniquely in children older than 6 years. It is frequently misdiagnosed, because of its rarity and its varied and atypical symptoms, but it may induce serious – even fatal complications, due to the involvement of many intracranial structures.

We report 3 cases of isolated sphenoiditis in previously healthy children, with various symptoms indicating acute central nervous system (CNS) disorder: A 4 year old boy with sommolence and meningitis-like symptoms, an 11 year old boy with visual illusions and a 12 year old boy with headache, emesis and positive Romberg's sign. In the initial clinical estimation there was no strong suspicion of infection, except in the 1st patient, because of the atypical symptoms. Diagnosis was established by a head CT scan, and all patients were treated with intravenous clindamycin plus cephalosporine, plus one dose methylprednisolone. The outcome was good.

Descriptors: SPHENOID SINUSITIS; CHILDREN; SIGNS AND SYMPTOMS

INTRODUCTION

Isolated sphenoid sinusitis is unusual (incidence about 2.7% of all sinus infections) and even rarer in children under 6 years old (1, 2). It is frequently misdiagnosed because of its atypical presentation, but it threatens serious and even fatal complications, due to the sphenoid's anatomical relationships with many intracranial structures (nerves, vessels etc) (3). We report on three patients with isolated sphenoiditis who were previously healthy, with acute symptoms in the CNS.

CASE REPORTS

Patient 1

A 4-year old boy presented complaining of somnolence during the morning. He had slept normally and had neither fever nor other signs of infection. His parents declared that he could not have reached any medicine and they did not witness any convulsions. His previous medical history was uneventful. On arrival he was somnolent, had difficulty responding to vocal stimuli, with no other focal neurological signs. Physical examination was normal. Investigations showed a raised white cell count of 29 000/mm³ (85% neutrophills, 9% lymphocytes) normal hemoglobin and platelets. C-reactive protein was 0.11 mg/dL and ESR 8 mm/1h. The other biochemical examinations, urea and urinalysis were within normal range. Cerebrospinal fluid examination was clear, gram stain and culture was negative. A CT scan of the head revealed left sphenoid sinus opacity, other sinuses and brain structures were normal (figure 1). Treatment with i. v. ceftriaxone and clindamycin was initiated. During the first hours of hospitalization he manifested symptoms indicating meningitis (intense headache, photophobia, emesis, stiff neck). A single dose of methylprednisolone was added, after an otorhinolaryngologist consultation, as a traditional local practice in complicated sinusitis. The next day the meningism subsided and the rest of his course was uneventful. He was treated with i. v. antibiotics for 10 days, then he was switched to oral cefuroxime axetil for 10 days. His final outcome was excellent.

Patient 2

An 11-year old boy presented with a 2-day feeling of dizziness and visual blurring. He also mentioned two episodes of visual illusions the previous day (he had twice seen animals near him, while he actually knew that they did not really exist). He had no fever, headache nor other signs of any infection. There was a history of minor head injury a month before with a headache, that did not require any medical intervention. The rest of his medical history was uneventful. Physical and neurological examinations were unremarkable. CBC, glucose, serum electrolytes, blood urea, creatinine and aminotransferases and urinalysis on admission were normal. Because of the head injury history a CT scan of the head was performed

* Giannitsa General Hospital- Pediatric Department, Giannitsa, 58100, Greece

Correspondence to:
Monti Catherine, Grigoriou E 15, Kalamaria 55 133, Thessaloniki, Greece, e-mail: monthcon@otenet.gr

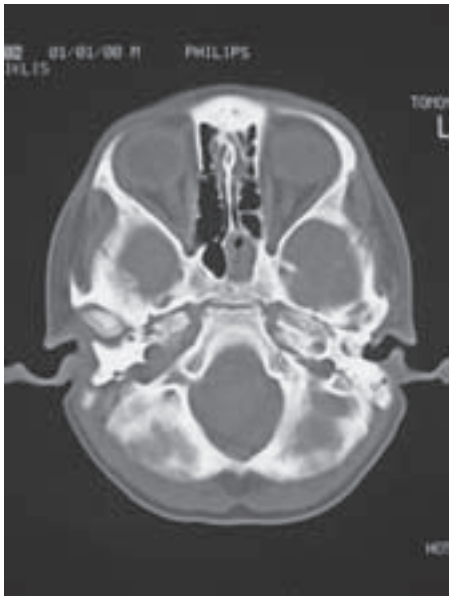

Figure 1. *Left sphenoid sinus opacity*
Slika 1. *Zasjenjenje u području lijevog sfenoidnog sinusa*

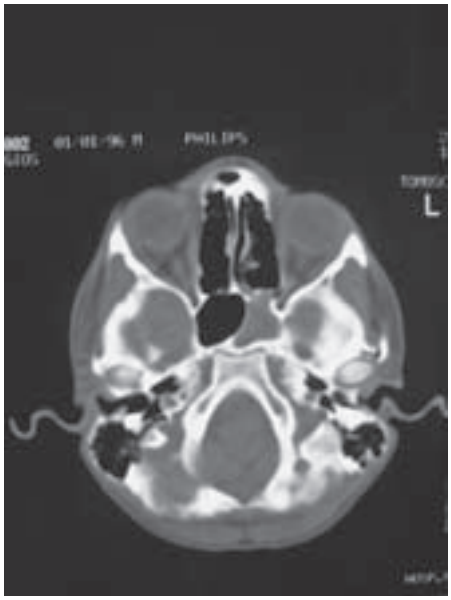

Figure 2. *Complete opacification of left sphenoid sinus*
Slika 2. *Potpuno zasjenjenje u području lijevog sfenoidnog sinusa*

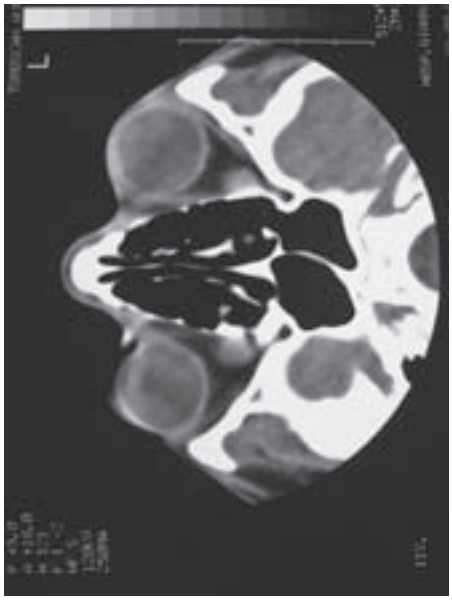

Figure 3. *Imaging after treatment shows full bilateral pneumatosis*
Slika 3. *Nakon provedenog liječenja – sinusi u potpunosti pneumatizirani*

and revealed total opacification of the left sphenoid sinus (figure 2). Clindamycin plus cefuroxime treatment was initiated i. v., plus methylprednisolone (for 1 day). He had an uneventful hospitalization, except an episode of vision blurring (a few hours after admission). In order to investigate any possibility of psychic origin of his symptoms, psychiatric examination and follow-up were made and excluded that suspicion. He was treated with i. v. antibiotics for 10 days and was given a cefuroxime axetil *per os* for 10 more days. The patient was discharged in good condition and a repeat CT scan after 10 days showed full regression of the sphenoid sinusitis (figure 3).

Patient 3

A 12-year old boy presented with acute and severe headache, dizziness and emesis. The headache was diffuse, frontal in location and constant in nature. He was without any previous history of headaches or migraine. The rest of his medical history was also uneventful. On physical examination, he had normal vital signs and mental status but during a more thorough neurological observation showed a positive Romberg's sign. Laboratory findings of blood and urine were normal. A CT scan of the head was performed and it revealed chronic right sphenoid sinusitis (figure 4). He was treated with intravenous ceftriaxone plus clindamycin i. v. for 10 days, in combination with methylprednisolone on the first day, and a course of cefuroxime axetil plus clindamycin *per os* was prescribed (which he finally discontinued at day 7). At a follow up examination after 15 days, he was in good condition but the CT scan showed no improvement. Any fungal origin was excluded radiologically, as well as mucocele or tumor, and the finding was diagnosed as a retention cyst. According to the otorhinolaryngologist, no surgical approach was necessary, and nasal steroid spray was prescribed for at least 3 months to facilitate drainage. At follow up visit after 6 months the boy was in excellent health and did not complain of any headache.

DISCUSSION

Sphenoid sinusitis is identified in approximately 3% of patients with acute sinusitis, typically in the context of pansinusitis (1). The sphenoid sinus is lined with ciliated pseudostratified epithelium with fewer mucous secreting cells as compared to the other paranasal sinuses. This contributes to fewer drainage problems and may explain the low incidence of isolated sphenoiditis (2). Significant development of the sphenoid sinuses does not begin until age 4-6 years, thus, sphenoid sinusitis is restricted to older children and adolescents. Likewise, intracranial complications of sinusitis appear predominantly in older children (4). Our 4 year old first patient was an especially rare case. Approximately 80% of paediatric patients with intracranial complications of sinusitis have sphenoid involvement (5).

Various predisposing factors for acute sphenoiditis have been identified. These include anatomical obstructions such as abnormally placed or small sphenoid ostiums, septal deviation and large superior or middle turbinates (6). Injuries have been shown to lead to infection, swimming and diving have also been linked to acute sphenoiditis (7). Other predisposing factors could be sphenoid ostium obstruction secondary to mucosal oedema and allergic rhinitis (7). None of our patients had recognizable predisposing factors.

The anatomic location of the sphenoid sinus places it adjacent to the optic canals, dura mater, cavernous sinuses, cranial nerves III to VI, and the internal carotid arteries. The bony walls of the sphenoid are thin or sometimes absent, leaving the sinus separated from intracranial structures by a narrow mucosal barrier (8). Any of the structures related to the sphenoid sinus can be affected by pathological processes involving the sinus. Proetz listed 13 structures adjacent to the sphenoid sinus that may be affected by disease: cranial nerves II, III, IV, cranial nerves V1, cranial nerves V2, cranial nerves VI, the dura mater, pituitary gland, cavernous sinus, internal carotid artery, sphenopa-

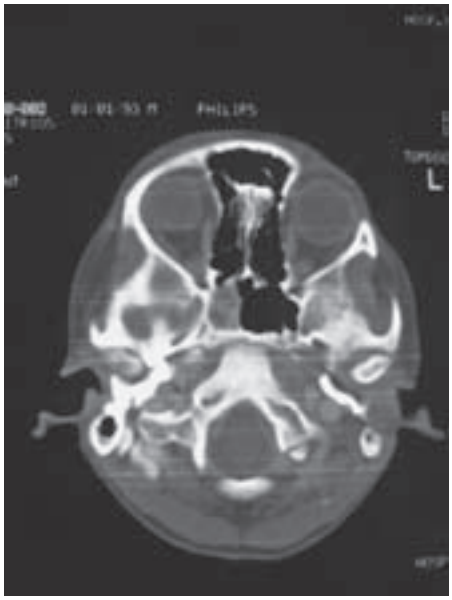

Figure 4. *Right sphenoid sinusitis radiologically estimated as chronic*
Slika 4. *Sinusitis desnog sfenoidnog sinusa - radiološki slika kronične upale*

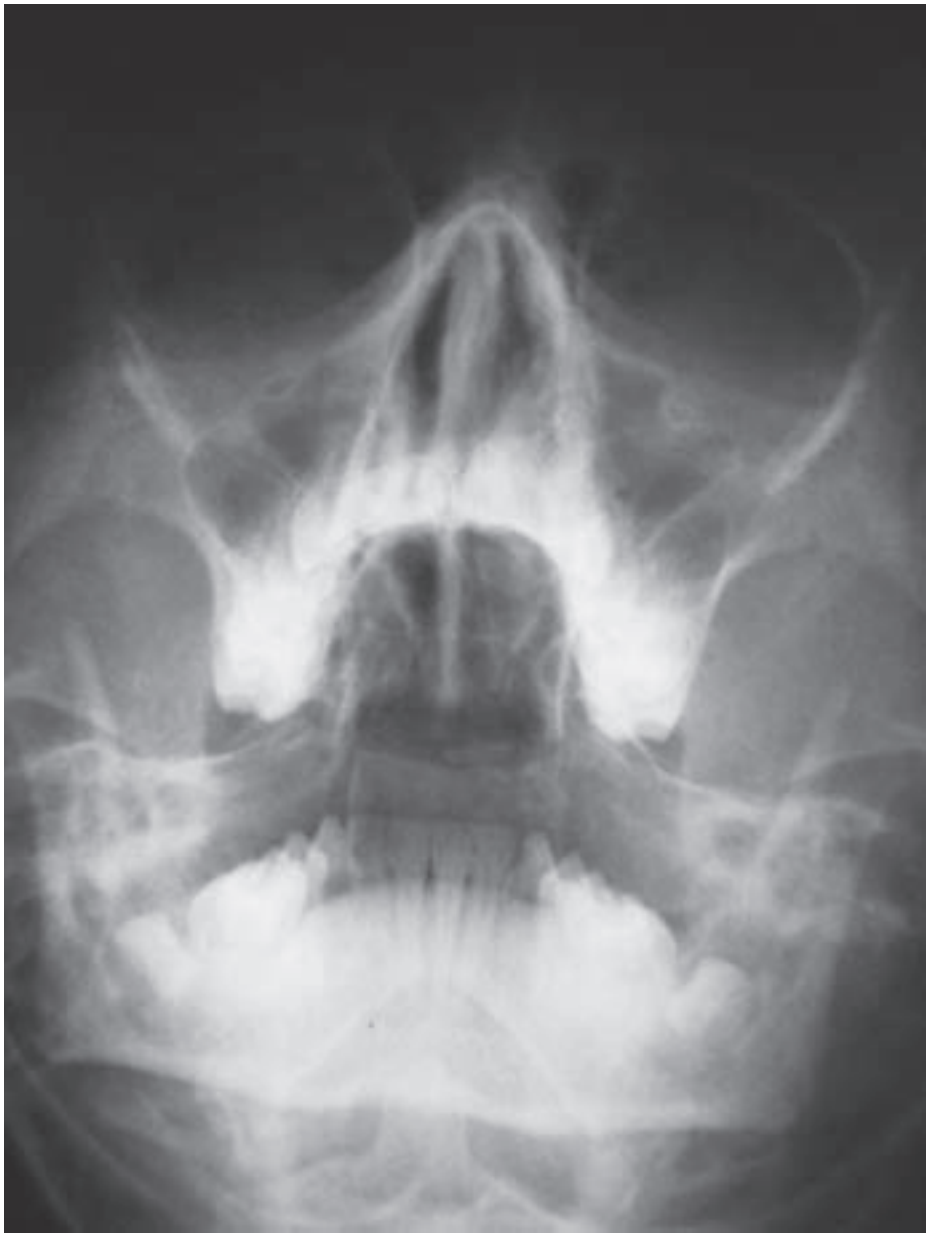

Figure 5. *An X-ray plan of sinuses in a 8-year old child: simple opacity of the left sphenoid*
Slika 5. *Rengenska snimka sinusa u 8-godišnjeg djeteta – jednostavno zasjenjenje u području lijevog sfenoidnog sinusa*

latine ganglion, sphenopalatine artery, pterygoid canal and nerve (3).

Sphenoid sinusitis can thus lead to orbital cellulites and abscess, orbital apex syndrome, blindness, sepsis, meningitis, epidural and subdural abscess, cerebral infraction, pituitary abscess, cavernous sinus and internal carotid artery thrombosis (9). For our second patient, the transient episodes of visual illusions were probably due to optic nerve involvement, with ischemia being the most probable mechanism in certain cases (10). Our 1st patient had symptoms of meningitis. It is hypothesized that sphenoid sinus infection spreads to the meninges by direct penetration of the sinus wall or by retrograde extension along the valveless diploic veins. Alternatively, a systemic bacteraemia could arise with subsequent penetration of the blood-brain barrier (4). In our patient, lumbar puncture showed normal cerebrospinal fluid (CSF). Consequently, we hypothesize that direct penetration to the meninges from the sinus wall was the primary event in this case.

Headache is the most common initial symptom of acute sphenoiditis (7, 9, 11). This was the presenting complaint in our third patient. Headaches have been described in descending order of frequency as deep-seated retro-orbital, frontal over the vertex, temporal, occipital or postauricular (12). They are more often non-specific and may present anywhere in the craniofacial region (13). Facial pain is thought to be due to the involvement of V1 and V2 nerves. In our patient the headache was diffusely frontal, non-specific and constant. His positive Romberg's sign was initially interpreted as a sign of raised intracranial pressure, not related to infection. In first patient the initial symptom was somnolence while headache manifested later with other signs of meningeal irritation.

The absence of nasal symptoms does not preclude the presence of sphenoiditis, as illustrated in all our patients. Significant physical findings are usually absent, though the presence of neurological findings would suggest an intracranial complication. Our 1st patient manifested sings of meningitis a few hours after admission, in 3rd patient the neurological examination showed a positive Romberg's sign and 2nd patient had the unusual symptom of visual illusions.

A 78% rate of misdiagnosed cases is reported in literature, attributed to atypical presentation (14). The diagnostic study of choice is a high resolution CT scan (axial and coronal views). This may reveal the presence of fluid or opacification and delineate the walls of the sphe-

noid sinus. In sphenoid sinusitis there is usually no bony erosion as compared to malignant disease. MR imaging with contrast should be obtained if there are any cranial nerve abnormalities as it is superior to CT scan in revealing pathology in the cavernous sinus and its adjacent neurovascular structures (15). Routine radiographs usually fail to reveal sphenoiditis, because of its deep location. Sphenoid opacity alone on plain X-rays (figure 5) is not diagnostic of sinusitis, except when air or fluid levels are present.

The most common pathogen in sphenoiditis is *Staphylococcus aureus*, followed by streptococcal species (1, 16). Fungi, especially *Aspergillus*, must be considered in all patients particularly if the patient is immunocompromised (6). Gram-negative and anaerobic organisms have been occasionally cultured. *S. pneumoniae* appears to be the pathogen most frequently associated with the development of meningitis as a complication (4).

Uncomplicated cases of acute sphenoiditis can resolve with optimal antibiotic therapy if diagnosed and treated early (1, 9). Prompt treatment is necessary as delay can result in serious morbidity and mortality (1, 16). The choice of antibiotics should take into account the wide spectrum of organisms isolated, and these include a combination of high-dose clindamycin (targets *S.aureus, Streptococcus species* and anaerobes) and a third generation cephalosporin (targets gram-negative organisms) given parenterally (17). Patients who are allergic to cephalosporin can be given aminoglycosides instead. Cloxacillin can be added for additional cover for *S.aureus* (17). Specific antimicrobial therapy can be adjusted once the culture results from the CSF, blood and sinus aspirates are known. The duration of antibiotic treatment is about 3 to 4 weeks (16). Topical decongestant and saline irrigation help to promote drainage of the obstructed sinus. In our first patient the CSF culture was negative and in all 3 cases therapy was given empirically based on the most common pathogens, for about 3 weeks (10 days parenterally and 10 days orally). Methylprednisolone was added for one day after consultation with otolaryngologist, for its anti-inflammatory and mucolytic effects, as a traditional therapy of our Hospital's ENT Department practice. Although similar references can be found, especially when referring to chronic cases, solid evidence for systemic steroids in sphenoiditis does not exist and remains a controversial issue (18). If symptoms progress or continue for more than 24 hours, or if complications arise, immediate surgical drainage of the sphenoid sinus is indicated (6). Various approaches to the sphenoid sinus are available. None of our patients needed surgical drainage of the sphenoid.

CONCLUSION

Sphenoiditis is a rare infection, even rarer in young children. Its symptoms are usually atypical and varied, making the diagnosis difficult or false. We reported 3 paediatric patients with isolated sphenoiditis, manifested by neurological symptoms that could easily be misdiagnosed without CNS imaging. Clinicians have to adopt a high level of suspicion in young children with serious or unexplained symptoms involving the central nervous system, and consider imaging for early diagnosis of potentially life threatening situations.

REFERENCES

1. Lew D, Southwick FS, Montgomery WW, Weber AL, Baker AS. Sphenoid sinusitis. A review of 30 cases. N Engl J Med 1983;309:1149-54.
2. Maran AGD, Lund VJ. Clinical Rhinology. New York: Thieme Medical Publishers, 1990.
3. Proetz AW. The sphenoid sinus. Br Med J 1948; 2:243-5.
4. A. Saitoh, B. Beall, V. Nizet. Fulminant bacterial meningitis complicating sphenoid sinusitis. Pediatr Emerg Care 2003;19:415-7.
5. Giannoni C, Sulek M, Friedman EM. Intracranial complications of sinusitis: a pediatric series. Am J Rhinol 1998;12:173-8.
6. Van Alyea OE. Sphenoid sinus: anatomic study, with consideration of the clinical significance of the structural characteristics of the sphenoid sinus. Arch Otolaryngol 1941;34:225-53.
7. Lahat E, Danieli J, Cohen H, Paret G, Barzilai A. Acute isolated sphenoid sinusitis in children. Pediatr Infect Dis J 1997;16:1180-2.
8. Levine H. The sphenoid sinus, the neglected nasal sinus. Arch Otolaryngol 1978;104:585-7.
9. Urquhart AC, Fung G, McIntoch WA. Isolated sphenoiditis: a diagnostic problem. J Laryngol Otol 1989;103:526-7.
10. Berlucchi M, Rossinia M, Bondioni M, Nicolai P. Isolated acute sphenoiditis with visual loss: A rare disorder in pediatric patients. Int J Pediatr Otorhinolaryngol Extra 2008;3:132-13.
11. Holt GR, Standefer JA, Brown WE Jr, Gates GA. Infectious diseases of the sphenoid sinus. Laryngoscope 1984;94:330-5.
12. Pearlman SJ, Lawson W, Biller HF, Friedman WH, Potter GD. Isolated sphenoid sinus disease. Laryngoscope 1989;99:716-20.
13. Lawson W, Reino AJ. Isolated sphenoid sinus disease: an analysis of 132 cases. Laryngoscope 1997;107:1590-5.
14. Hnatuk LA, Macdonald RE, Papsin BC. Isolated sphenoid sinusitis: the Toronto Hospital for Sick Children experience and review of the literature. J Otolaryngol 1994;23:36-41.
15. HKK Tan, FRCS,YK Ong. Acute isolated sphenoid sinusitis. Ann Acad Med Singapore 2004;33: 656-9.
16. Haimi-Cohen Y, Amir J, Zeharia A, Danziger Y, Ziv N, Mimouni M. Isolated sphenoidal sinusitis in children. Eur J Pediatr 1999;158:298-301.
17. Brook I. Bacteriology of acute and chronic sphenoid sinusitis. Ann Otol Rhinol Laryngol 2002; 111:1002-4.
18. Cable BB, Wassmuth Z, Mann EA, Hommer D, Connely G, Klem C, Quance-Fitch FJ, Bolger WE. The effect of corticosteroids in the treatment of experimental sinusitis. Am J Rhinol 2000;14:217-22.

S a ž e t a k

NEUROLOŠKI SIMPTOMI KAO POSLJEDICA IZOLIRANOG SFENOIDALNOG SINUSITISA U DJECE: PRIKAZ TRIJU SLUČAJEVA

M. Farini, S. Totska, C. Monti

Upala sfenoidnog sinusa je rijetka, a najčešće nastaje u sklopu infekcija ostalih paranazalnih sinusa. Izolirani sfenoiditis još je rjeđi, a opisuje se gotovo uvijek u djece starije od 6 godina. Često ostaje neprepoznat, jer može davati različite i nerijetko atipične simptome, a također, zbog blizine mnogih intrakranijalnih struktura, može rezultirati ozbiljnim ili smrtonosnim komplikacijama.

Prikazujemo troje bolesnika s izoliranim sfenoiditisom, koji su prethodno bili zdravi, a očitovali su se različitim simptomima koji su upućivali na oštećenje središnjeg živčanog sustava. Četverogodišnji dječak je bio somnolentan i imao simptome slične meningitisu, 11-godišnji dječak je imao vizualne halucinacije, a 12-godišnjak je imao glavobolju, povraćanje i pozitivni Rombergov znak. Pri inicijalnom pregledu, zbog atipičnih simptoma, nije se sumnjalo na infekt, osim u prvog dječaka. Dijagnoza je postavljena uz pomoć CT-a, a sva djeca su liječena kombinacijom intravenskog klindamicina i cefalosporina i jednom dozom metilpredinzolona. Ishod liječenja je bio dobar.

Deskriptori: SFENOIDNI SINUSITIS; DJECA; ZNAKOVI I SIMPTOMI

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