Calvarial involvement in disseminated rhinosporidiosis – A case report and literature review

Dewangan B, Naik R, Membally R, Dewangan M

ABSTRACT

Rhinosporidiosis is a chronic granulomatous infection caused by *Rhinosporidium seeberi* and mainly involves nasal and ocular mucosa. Bony involvement in rhinosporidiosis is very rare. A young male, previously operated for nasal rhinosporidiosis, presented with two bony swellings on the forehead and multiple subcutaneous lesions on the right lower limb. The diagnosis of disseminated cutaneous rhinosporidiosis with frontal bone involvement was made with the help of fine needle aspiration cytology (FNAC), histopathology, and computed tomography (CT) scan head. Wide excision of the bony lesion was performed. To the best of our knowledge, this is the first radiologically proven case of frontal bone involvement in disseminated rhinosporidiosis. Early diagnosis can be established with a good clinicopathological and radiological correlation. It also emphasizes the importance of CT scan for the evaluation of any subcutaneous skull lesion.

KEY WORDS: Calvarial rhinosporidiosis, disseminated cutaneous rhinosporidiosis, frontal bone rhinosporidiosis, osseous rhinosporidiosis

Introduction

*Rhinosporidiosis* is caused by *Rhinosporidium seeberi*, an aquatic protistan parasite of the *Mesomycetozoea* class. It usually involves nasal mucosa and conjunctiva, but mucous membranes at other sites may also be affected. Osseous involvement is a very rare occurrence in this disease. Calvarial involvement in disseminated rhinosporidiosis is not described in the literature. We present here the first radiologically as well as histopathologically proven case of frontal bone involvement in disseminated rhinosporidiosis.

Case Report

A 27-year-old man from Chhattisgarh presented with painless swelling over the right side of the forehead since 6 months and multiple swellings on the right leg since 2 months. He had undergone excision of a nasal mass 5 years back in a private hospital and histopathology from the excised lesion was not done. He was a farmer and a regular user of the village pond for bathing which was also shared by cattle.

Physical examination revealed two well-defined hard, nontender subcutaneous swellings over the right side of forehead just above the eyebrow varying from 5 × 5 to 2 × 2 cm in size [Figure 1a]. There was central crackling sensation on one of the swellings. There were three lesions on the right leg. The two were well-defined subcutaneous nodules − 3 × 4 and 2 × 3 cm on the medial aspect and one was diffuse cystic swelling on the lateral aspect [Figure 1b]. History of shared pond bathing and surgery for nasal mass excision in the past raised the suspicion of disseminated rhinosporidiosis. Fine needle aspiration of all the swellings was suggestive of rhinosporidiosis [Figure 2a]. Roentgenography of the skull and lower limbs appeared normal. Computed tomography (CT) scan of the head showed erosion of the frontal bone at two corresponding sites [Figure 3a-f].
The final diagnosis of disseminated cutaneous rhinosporidiosis with frontal bone involvement was made. Tablet dapsone 100 mg daily was started, but there was no change after 2 months of therapy. In fact, the size of his forehead lesion has gradually increased to cover the upper half of the orbit and eye opening was restricted. Surgical management was planned. The two subcutaneous nodules of leg were subjected to excisional biopsy along with electrocoagulation of the margins. Aspiration of the diffuse swelling on the lateral aspect of right leg yielded serosanguinous fluid and was left after decompression for further observation.

On exploration of the forehead lesion, there was fragile granulation tissue with serosanguinous fluid over the eroded area of the frontal bone. Fluid was drained and the granulation tissue was curetted out. The content was sent for histopathological examination which confirmed rhinosporidiosis. Both outer and inner table of the frontal bone was deficient in the bigger lesion whereas inner table was intact in the smaller one [Figure 4]. Dura was not breached in either cases. One centimeter margin of the bone was excised all around and subjected for histopathological examination. Histology confirmed the involvement of bone with the presence of sporangia in the marrow space [Figure 2b]. Postoperative recovery was uneventful and he was discharged on tablet dapsone 100 mg once daily. After 1 year of follow-up, there are no signs of recurrence [Figure 1c].

**Discussion**

Rhinosporidiosis has been reported from all over the world but it is more common in tropics. The disease prevalence is high in some parts of India and Sri Lanka.\(^1\) In India, the disease is mainly confined to the coastal states of south India where prevalence is as high as 4.7% and Chhattisgarh region with a prevalence rate of 1%.\(^2\)

Four forms of the rhinosporidiosis are recognized as per the anatomical localization – nasopharyngeal (70%–90%), ocular (15%), cutaneous, and disseminated.\(^3\) Cutaneous and systemic dissemination is quite rare.\(^3\) Occasionally lips, palate, uvula, maxillary antrum, epiglottis, pharynx, larynx, trachea, bronchus, ears, vulva, vagina, urethra, penis, rectum, scalp, and skin are involved.\(^3,4\) Bony involvement is extremely rare. Only 18 cases have been reported so far in the literature till date, but frontal bone involvement has been reported once only [Table 1].\(^4,20\)

Bone involvement in rhinosporidiosis can occur in three different clinical settings: 1) Local invasion of the skull bone by nasal rhinosporidiosis. 2) Bone involvement is a part of disseminated rhinosporidiosis as seen in our case. 3) Bone involvement is primary without any evidence of lesions elsewhere.\(^4\) Hematogenous spread could be the possible mode of spread to the distant sites.\(^21\) Frontal bone, as in our case could have been reached by this route only.

The sole previous report by Madhavan et al. described isolated frontal bone involvement in 1978, but the radiological evidence was lacking and the diagnosis of osseous involvement was made postoperatively after histopathological examination.\(^6\) Our case is unique because the frontal bone affliction was the part of disseminated rhinosporidiosis and was radiologically evident. Furthermore, the frontal bone erosion was not depicted by the routine X-ray. It was revealed only on CT scan. Hence, we recommend CT scan for the evaluation of any skull lesion in disseminated cutaneous rhinosporidiosis to detect underlying bony involvement.

CT imaging features of surrounding bony involvement in rhinosporidiosis has been described as irregularity, rarefaction, complete, or partial erosion.\(^22\) The sunburst or spiculated pattern of periosteal reaction as seen in our case has not been described in the literature previously. These CT imaging features can easily be confused with bony tumors such as osteosarcoma, multiple myeloma, and metastasis from unknown primary. Therefore, a good clinicopathological and radiological correlation is necessary to diagnose calvarial rhinosporidiosis preoperatively.

Treatment of choice is excision and electrocoagulation of the base of subcutaneous lesion and wide excision of the bony
Table 1: Description of all the cases of osseous involvement in rhinosporidiosis described in literature with respect to involved bone, associated lesions and treatment

| Serial no | Author | Bones involved | Associated lesions | Treatment of bony lesion |
|-----------|--------|----------------|--------------------|--------------------------|
| 1         | Chatterjee et al., 1977[5] | 1. Proximal phalynx of the left ring finger 2. Second metatarsal, proximal and middle phalynx of second right toe | Mass in the nose, nodules in the face, left axilla, left chest wall and calf | Not described |
| 2         | Madhvan M et al., 1978[6] | Right frontal bone | Nil | Curettage |
| 3         | Sudarshan et al., 1979[7] | First metatarsal of left foot | Nodule in nasopharynx | Curettage |
| 4         | Aravindan et al., 1989[8] | Scapula | Nil | Excision |
| 5         | Mitra and Maity et al. 1996[9] | Right calcaneum | Nil | Not described |
| 6         | Pai S.A. et al. 1996[10] | Fifth metacarpal, proximal phalynx of the right ring finger | Nil | Not described |
| 7         | Adiga B K, et al. 1997[11] | Tibia | Nil | Wide excision |
| 8         | Gokhale S. et al. 1997[12] | Proximal phalynx of fourth and base of fourth and fifth Metacarpal | Nodule in scalp, mass over third toe | Partial amputation of hand |
| 9         | Kavishwar VS et al. 1998[13] | Fourth and fifth metacarpals | Nasal polyp | Not described |
| 10        | Makannavar JH et al. 2001[14] | Tibial condyle | Nasal polyp | Not described |
| 11        | Dash et al. 2005[15] | Femur | Not available | Not described |
| 12        | Sudarshan V et al. 2007[16] | Not described | Not described | Not described |
| 13        | Amritanand R et al. 2008[17] | Talus, calcaneum, tarsals and bases of first and second metatarsals | Warts on face | Below knee amputation |
| 14        | Suryavanshi P V et al. 2011[18] | Clavicle | Nil | Wide excision |
| 15        | Mondal et al. 2013[19] | Left distal end radius and ulna, all carpals and base of metacarpals | Nil | Below elbow amputation left side |
| 16        | Kundu AK, et al. 2013[20] | Fifth metacarpal Right side | Poly in nose and lachrymal gland | Ray amputation |
| 17        | Pal D K et al. 2013[21] | Lower end of radius and carpal bones | Nil | Wide excision |
| 18        | Acharya S et al. 2014[22] | Calcaneum | Nasal mass, nodule over right forearm | Not described |

Figure 3: Transaxial enhanced computed tomography images of the brain. (a) Right supraorbital homogenous enhancing soft tissue swelling in subcutaneous plane (arrow). (b) Bone window showing adjacent irregular osseous destruction (arrow) extending up to inner table and lateral wall of right frontal sinus. (c) Enhancing soft tissue swelling extending to right frontal extracalvarial region (arrow). (d) Bone window shows mild osseous erosion of the outer table with thick solid spiculated periosteal reaction (sunburst appearance) (arrow). (e) Small focal enhancing lobular soft tissue in midline high frontal subcutaneous plane (arrow). (f) Bone window showing adjacent punched out osteolytic destruction (arrow).
To conclude, almost all the bones are within the reach of *R. seeberi*, probably through the hematogenous route. It produces osteolytic lesions mimicking various bony lesions including tumors. High index of suspicion will pick up the lesions early, especially in areas with high prevalence. Diagnosis can easily be confirmed by FNAC and histopathology. Excision of the lesion as wide as possible is the treatment of choice. Tablet dapsone is recommended to prevent recurrence.

**Declaration of patient consent**
The authors certify that appropriate patient consent was obtained.

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Nil.

**Conflicts of interest**
There are no conflicts of interest.

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