CASE REPORT

Rhinofacial entomophthoromycosis

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Entomophthoromycosis is a rare subcutaneous fungal infection caused by *onidiobolus coronatus* affecting mainly the upper respiratory mucosa in immunocompetent people. The manifestations of this disease masquerades other clinical entities. Hence, high index of suspicion is required for prompt diagnosis. Histopathological examination and culture are the gold standard diagnostic tools, however no standard treatment protocols have been mentioned in literature. We present a case of rhinofacial entomophthoromycosis in a yearold ma with a leftsided rhinofacial swelling to highlight the presence of this unusual fungal infection and its treatment.

Figure 2 Rhinofacial swelling.

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BACKGROUND

Rhinofacial entomophthoromycosis, also known as conidiobolomycosis, is a rare subcutaneous zygomycosis that is caused by *Conidiobolus coronatus*. It is seen mainly in the tropical and subtropical regions of the world. It primarily affects the upper respiratory mucosa and adjacent subcutaneous tissue in immunocompetent people. It is known to infect nasal mucosa and submucosa forming subcutaneous nodules with the potential to spread to adjacent tissues. Because of this localised spread, it is known as rhinofacial entomophthoromycosis.¹ It can be mistaken for malignancy or tuberculosis. It is seldom life-threatening but may cause severe facial disfigurement. The diagnosis is based on a combination of tissue biopsy, culture and clinical presentation.

CASE PRESENTATION

A 30-year-old man presented with a left nasofacial swelling and left-sided nasal obstruction of 3 months' duration. On examination, a firm-to-hard subcutaneous swelling was seen in the upper gingivolabial sulcus extending to the floor and lateral ala of the nose on the left side. Posteriorly, it was extending until the inferior turbinate and the

adjoining maxillary sinus. The lesion was causing prominence of the philtrum, upper lip and columella. There was obliteration of the nasolabial and nasomaxillary creases, going onto the frontal process of maxilla just short of medial canthus. The skin over the swelling was normal and pinchable (figures 1 and 2)

INVESTIGATIONS

A contrast-enhanced CT scan of the nose and paranasal sinuses showed a heterogeneously enhancing lesion involving the left nasal cavity involving the floor, lateral wall in the region of upper and lower lateral cartilage and adjoining inferior turbinate, extending on to the anterolateral wall of the maxilla. The extension superiorly was over the frontal process of the maxilla (figure 3). A diagnostic nasal endoscopy was done which showed a smooth submucosal swelling involving the floor and lateral wall of the nasal vestibule and the left inferior turbinate (figure 4). A biopsy was taken from the lesion under topical anaesthesia which showed features suggestive of zygomycosis.

DIFFERENTIAL DIAGNOSIS

Under general anaesthesia, using sublabial approach, the lesion was exposed and removed piecemeal by dissecting it free from the anterior part of the left nasal cavity. The adjoining involved part of the upper and lower lateral cartilage was excised. The involved left inferior turbinate was removed by partial turbinectomy (figure 5).

OUTCOME AND FOLLOW-UP

The histopathology of the specimen showed respiratory epithelium overlying fibrous tissue showing

Figure 1 Rhinofacial swelling (left)

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Figure 3 CT nose and paranasal sinus.

discrete granulomas with amorphous eosinophilic Splendore-Hoeppli material. The adjoining granuloma showed few thin-walled fungal hyphae (periodic acid–Schiff (PAS) positive). The diagnosis of rhinofacial entomophthoromycosis was made (figure 6).

The patient was planned for oral itraconazole 200 mg twice daily for 2 months. Due to deranged liver function test, itraconazole was discontinued after 6 weeks. On follow-up after 4 months and 1 year, there was no evidence of recurrence on clinical examination and nasal endoscopy. However, the patient had a deformity in the left alar region but no nasal obstruction.

DISCUSSION

C. coronatus is a very rare cause of nasal infection seen in immunocompetent people residing in the tropical and subtropical regions.^{1 2} *Conidiobolus* is a saprophytic fungus present in decaying vegetative matter and soil of the order entomophthorales of the class Zygomycetes.³ The infection has been suggested

to result from percutaneous inoculation of *C. coronatus* via inhalation of fungal spore which implants in the nasal mucosa or via an insect bite.^{2 3} Most of the infected people are known to be engaged in agricultural or an outdoor occupation. It is a slow-growing, locally infiltrative disease of the nasal cavity, paranasal sinuses and soft tissues of the face. The first to report a case of rhinofacial zygomycosis in humans was by Lie Kian Joe in Indonesia in 1960.³

The common clinical presentation is a subcutaneous nodular lesion presenting with nasal obstruction, nasal discharge, epistaxis and deformity.^{3 4} It may mimic a malignancy or a granulomatous disease like tuberculosis or angiocentric lymphoproliferative disorders.⁴ The bones are usually not involved but it can spread to adjacent paranasal sinuses and facial soft tissues. The spread to the mediastinum and lungs has also been reported. Hence, an early and accurate diagnosis is essential.⁵ It can also present with orbital extension leading to proptosis and loss of vision.⁶

Diagnosis is mainly by biopsy and tissue culture. Histologically, entomophthorales are characterised by a chronic granulomatous inflammatory reaction comprising lymphocytes, eosinophils, histiocytes and foreign body type of multinucleated giant cells. The central portion comprises few broad septate hyphae surrounded by an amorphous eosinophilic material called the Splendore–Hoeppli phenomenon characteristic of entomophthorales.^{7 8} To demonstrate the fungal hyphae, PAS stain has been found to be useful.⁷ In contrast to mucormycosis, there is no angioinvasion or vascular thrombosis. Our case of a male patient presenting with nasal obstruction and facial deformity conformed to the clinical picture of *C. coronatus* infection which was confirmed by histology. Only 15% of cases are positive on mycological culture.⁸ A histological confirmation is important for diagnosis.

The treatment of choice for rhinofacial entomophthoromycosis is surgical resection followed by antifungal medication.^{9 10} Antifungal therapy of rhinofacial entomophthorales has not been well defined because of the infrequent nature of the disease. Reports of use of potassium iodide, cotrimoxazole, amphotericin B, imidazoles and a combination of these agents have been reported with varying success. The efficacy of these drugs has not been studied. The duration and dosage of these drugs are not well defined in literature because of the rarity of the disease.^{10 11} Itraconazole has been noted to be effective and relatively safe.¹²

Figure 4 Diagnostic nasal endoscopy showing the submucosal swelling.

Figure 5 Surgical approach: sublabial.

Figure 6 Histopathological examination (HPE): granuloma with fungal hyphae.

Learning points

- Rare fungal infection seen in immunocompetent people.
- Diagnosis is by histopathological examination.
- Combined modality of treatment: surgical excision followed by antifungal therapy is required.
- Regular follow-up is required to look for recurrence.

Our patient was treated with itraconazole for 6 weeks and showed good results. Relapse is said to be common even after successful treatment which makes follow-up mandatory.

Contributors SM is the corresponding author who was involved in collecting the clinical data and drafting the article. SM is also the assistant surgeon involved in the management of the case. KP is the principal surgeon of the case and has reviewed

the article with necessary inputs. RK is the pathologist reporting the case. BR was involved in the overall management of the case and has reviewed the final article.

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