**Objectives**:

To present a rare case of basidiobolomycosis in a young, immuno-competent patient with orbital and sinus involvement.

**Materials & Methods:**

A 19-year-old female from Balochistan, currently residing in Karachi, presented at a public sector tertiary care hospital in the clinic with a two-week history of headache and progressive right eye ptosis with loss of vision for one week. Imaging revealed extensive sinus inflammatory disease with orbital extension and sphenoid sinus swelling. Endoscopic biopsy of the sphenoid sinus was sent for histopathological examination, and a fungal culture was sent to the clinical laboratory at Aga Khan University Hospital. Histopathological analysis showed fibroblastic proliferation with patchy dense lymphoplasmacytic infiltrates and multinucleated giant cells. Additionally, ribbon-like, non-septate fungal hyphae were observed. Fungal culture of the sinus tissue initially revealed a very fine growth of whitish mold. By days 3 to 7, colonies rapidly expanded, were flat, cerebriform, and powdery, transitioning from white to yellowish grey on SDA and SBA at 250C. Microscopy showed pauciseptate broad hyphae with abundant rounded structures with dual protrusions resembling a parrot’s beak, identified as Basidiobolus species, confirming basidiobolomycosis.

**Results**:

The patient was given voriconazole 200 mg twelve-hourly, resulting in significant resolution of swelling. She was discharged with a recommendation to continue oral voriconazole for an additional two weeks and follow up in the clinic after 2 weeks.

**Conclusions**:

Basidiobolomycosis remains a diagnostic challenge due to its rarity and nonspecific clinical presentation. This case highlights the importance of considering Entomophthorales as a differential in chronic sinus infections with orbital extension and utilizing both fungal culture and histopathology for early identification. It serves as a compelling reminder of the need for heightened awareness and vigilance among clinicians regarding rare fungal infections, particularly in immunocompetent hosts, to ensure timely diagnosis and effective management. In light of the increasing global burden of emerging fungal pathogens, it is crucial to advance our diagnostic capabilities and broaden our understanding of these enigmatic infections.