Indian Journal of Medical Case Reports ISSN: 2319–3832(Online)
An Open Access, Online International Journal Available at http://www. cibtech.org/jcr.htm
2014 Vol. 3 (3) July-September, pp. 47-50/Selvi and Gulia
Case Report

POSTOPERATIVE DIAGNOSIS OF SUBCUTANEOUS PHAEOHYPHOMYCOSIS – A REPORT OF TWO CASES

***Kalaivani Selvi S. and Sandhya Panjeta Gulia**
Department Of Pathology, Sri Venkateswaraa Medical College Hospital And Research Centre (Affiliated to Pondicherry University), Puducherry, India
**Author for Correspondence*

ABSTRACT
Noonan syndrome [NS] is an autosomal dominant inherited condition that can be passed down through families. The incidence of NS is estimated to be between 1:1000 and 1:2500 live births. It causes abnormal development in many parts of the body. It is used to be called Turner-like syndrome. Though most of the cases are autosomally inherited some cases may be sporadic. We report a case of 11 year old male child presented to the hospital with features of cardiac failure and morphologic features of NS who has no similar history in the family.

Keywords: *Noonan Syndrome; Sporadic, Turner Like Syndrome*

INTRODUCTION
Phaeohyphomycosis is caused by a number of dematiaceous fungi where the tissue morphology of the causative organism is mycelial. This separates it from other clinical types of disease involving brown-pigmented fungi where the tissue morphology of the organism is a grain (mycotic mycetoma) or sclerotic body (chromoblastomycosis) (Revankar, 2007). Clinical forms of phaeohyphomycosis range from localized superficial infections of the stratum corneum (tinea nigra) to subcutaneous cysts (phaeomycotic cyst) to invasion of the brain and other systemic organs (McGinnis, 1983), Subcutaneous phaeohyphomycosis is the most common type (Saha, 2005) and is most frequently an opportunistic infection in immunosuppressed patients or associated with chronic diseases and diabetes (Parente, 2011) .

CASES
Case 1
A 65 year old male patient, resident ; a known case of hypertension for 10 years on medication presented with swelling near the elbow joint for three years. On examination was found to have a cystic swelling measuring 4x3 cm well defined with smooth surface, soft and mobile and was clinically diagnosed as a case of sebaceous cyst. Excision biopsy was done. On gross examination it was a cystic nodular mass with outer smooth surface measuring 4x3x3cm. Cut surface of the mass was uniloculated cyst filled with yellowish pultaceous like material with no solid areas (Figure 1a). Histopathological examination (HPE) showed fibrocollagenous cyst wall with granulomatous reaction with presence of histiocytes, mulitinucleated giant cells, lymphocytes, plasma cells with areas of fibrinoid necrosis admixed with numerous neutrophils in the cyst cavity (Figure 1b). Careful examination revealed brown pigmented septate fungal hyphae morphologically consistent with phaeohypomycosis (Figure 1c). PAS stain highlighted the fungal organisms (Figure 1d).

Case 2
A 49 year old female a known case of diabetic and hypertensive for 3 years presented with swelling in the right lateral malleolus for 3 years. On examination was found to have a globular smooth swelling measuring 4x3 cm. A clinical differential diagnosis of adventitious bursitis and lipoma was given. Excision biopsy was done. On gross examination it was a nodular, capsulated mass measuring 5x3.5x1.5 cms. Cut surface was multinodular with yellow pultaceous like material separated by fibrous septae (Figure 2a). HPE revealed fibrocollagenous capsule with large areas of fibrinoid necrosis and neutrophilic infiltration surrounded by granulommatous inflammation (Figure 2b) with brown pigmented fungal

organisms morphologically those of pheohyphomycosis (Figure 2c). PAS stain highlighted the fungal organisms (Figure 2d).

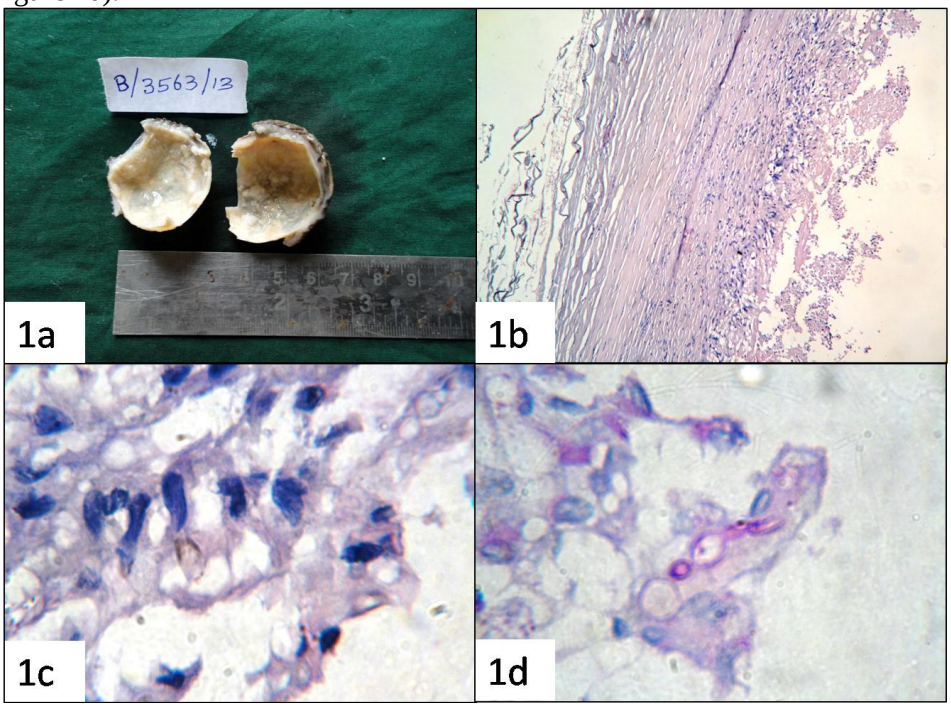

Figure 1: (a)Uniloculated cyst filled with necrotic material;(b)Fibrocollagenous cystwall with granulomatous inflammation and necrosis (H&E, 100X);(c)Brown pigmented septate hypae (H&E, 1000X);(d)PAS stain highlighting the fungus in the multinucleated giant cells (PAS, 400X)

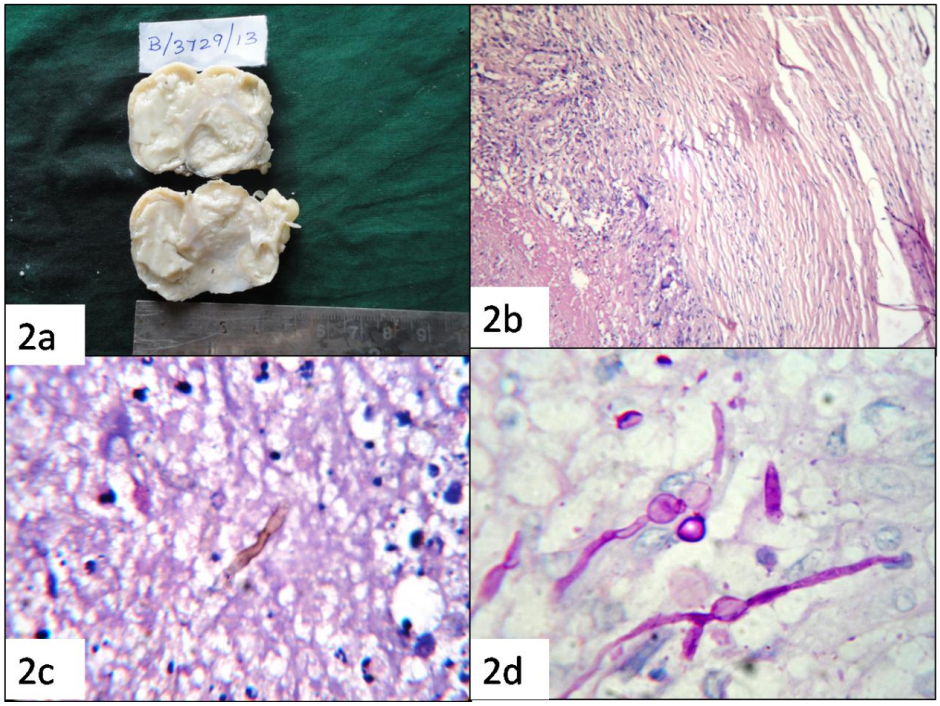

Figure 2: (a) Multinodular swelling filled with necrotic material;(b)Fibrocollagenous cystwall with granulomatous inflammation and necrosis (H&E, 100X);(c)Brown pigmented septate hypae (H&E, 400X);(d)PAS stain highlighting the fungus (PAS, 400X)

DISCUSSION

Phaeohyphomycosis is an umbrella term for all those mycoses where the fungus is present in host tissues with melanized filaments. The etiological agents include various species of *Exophiala, Phialophora, Wangiella, Bipolaris, Exserohilum, Cladophialophora, Phaeoannellomyces, Aureobasidium, Cladosporium, Curvularia* and *Alternaria.* (*Isa*-Isa, 2012). The etiologic agents of subcutaneous phaeohyphomycosis are predominantly Exophiala (E.) sp. and Phialophora (P) sp., including E. jeanselmei, E. dermatitidis and P. verrucosa (Kwon-Chung, 1992). The most common is E. jeanselmei, followed by E. dermatitidis (Saha, 2005; Kwon-Chung, 1992; Rinaldi, 1996).

The clinical presentation depends on the immune status of the host: superficial (tinea nigra and black piedra); cutaneous (scytalidiosis) and corneal; subcutaneous (mycotic cyst); and systemic phaeohyphomycosis in the immunocompromised host. Rarely, immunocompetent patients may be affected. The mycotic cyst is a localized form, characterized by subcutaneous asymptomatic nodular lesions that develop after traumatic implantation of fungus from contaminated soil, thorns or wood splinters, especially on the extremities (Saha, 2005; Kwon-Chung, 1992). The average size of the cysts is 2.5 cm (McGinnis, 1983).

Investigations include direct microscopy, culture and tissue examination. In Direct Microscopy using 10% KOH, the presence of brown pigmented, branching septate hyphae in any specimen, from a patient with supporting clinical symptoms should be considered significant. Biopsy and evidence of tissue invasion is of particular importance. Direct microscopy or histopathology does not offer a specific identification of the causative agent. It basically differentiates between chromoblastomycosis which is characterized by the presence of brown pigmented, planate-dividing, rounded sclerotic bodies and phaeohyphomycosis where the tissue morphology of the causative organism is mycelial. Culture identification is the only reliable means of identifying the specific fungal species.

Histopathology usually shows an abscess or a suppurative granuloma with brown septate hyphae or yeast or a combination of both in tissue. The Fontana-Masson stain, periodic acid-Schiff and Gomori methenamine-silver stains can be used to confirm the diagnosis (Revankar, 2007; Rinaldi, 1996). The treatment of choice is surgical excision, but additional anti-fungal therapy is recommended for recurrent cases and immunocompromised patients (Sharma, 2002).

Ajello listed 71 species from 39 genera as causative agents of phaeohyphomycosis (Ajello, 1974). The number of fungi documented as etiologic agents of phaeohyphomycosis currently number at least 57 genera and 104 species (Rinaldi, 1996).

In India so far only about 24 cases of subcutaneous phaeohypomycosis have been reported till 2010 (Pereira, 2010). Of the 24 cases 14 were males and 10 were females. Phialophora is the most common fungal organism isolated followed by cladosporium, curvilaria, exophiala, alterneria, fonsecaea and others. Extremities are more commonly involved, face, waist and buttock were other sites involved. Three cases were in disseminated form. The lesions were in the form of chronic ulcer, abscess, warty/verrucous plaques, cysts, nodules, scaly lesion and sinuses (Pereira, 2010; Sharma, 2002).

In this case report one was a male and the other was a female, presenting with subcutaneous cyst in the elbow and malleolus region with clinical diagnosis of sebaceous cyst and lipoma/bursitis respectively. On gross examination both the cases had pultaceous like material favouring sebaceous cyst. HPE showed a fibrous cyst wall, granulomatous reaction and pigmented fungi confirming the diagnosis of pheohyphomycosis.

The differential diagnosis with this particular gross finding and multinucleate giant cell reaction is ruptured epidermal inclusion cyst (EIC) with giant cell reaction and tuberculosis (TB bursitis). The absence of stratified squamous epithelium and presence of pigmented fungi rules out the diagnosis of EIC and TB in both the cases. Species identification by fungal culture could not be done in both cases as the tissue was already fixed in formalin.

In the absence of clinical suspicion, postoperative histopathological examination played a key role in diagnosis of subcutaneous phaeohypomycosis in both the cases.

Conclusion

Phaeohyphomycosis is a rare fungal infection and is even rarer in immunocompetent individuals. Any subcutaneous cystic swelling with granulommatous inflammation and necrosis with fibrocollagenous cyst wall in suspicious individuals should be carefully searched for pigmented fungal organisms to rule out subcutaneous phaeohyphomycosis.

REFERENCES

Ajello L, Georg LK, Steigbigel RT and Wang CJK (1974). A case of phaeohyphomycosis caused by a new species of Phialophora. *Mycologia* **66** 490-498.

Isa-Isa R, García C, Isa M and Arenas R (2012). Subcutaneous phaeohyphomycosis (mycotic cyst). *Clinics in Dermatology* **30**(4) 425-31.

Kwon-Chung KJ and Bennet JE (1992). Phaeohyphomycosis (chromomycosis, phaeosporotrichosis, cerebral chromomycosis). In: *Medical Mycology,* edited by Lea and Feblger, Philadelphia 620- 677.

McGinnis MR (1983). Chromoblastomycosis and phaeohyphomycosis: new concepts, diagnosis, and mycology. *Journal of the American Acadamy of Dermatology* **8** 1-16.

Parente JN, Talhari C, Ginter-Hanselmayer G, Schettini AP, Eiras Jda C, de Souza JV, Tavares R, Buzina W, Brunasso AM and Massone C (2011). Subcutaneous phaeohyphomycosis in immunocompetent patients: two new cases caused by Exophiala jeanselmei and Cladophialophora carrionii. *Mycoses* **54**(3) 265-9.

Pereira RR, Nayak CS, Deshpande SD, Bhatt KD, Khatu SS and Dhurat RS (2010). Subcutaneous phaeohyphomycosis caused by cladophialophora boppii. *Indian Journal of Dermatology Venereology and Leprology* **76** 695-8.

Revankar SG (2007). Dematiaceous fungi. *Mycoses* **50** 91-101.

Rinaldi MG (1996). Phaeohyphomycosis. *Dermatologic Clinics* **14** 147-153.

Saha R and **Rudra S (2005).** Phaeomycotic cyst--a case report. *Journal of Indian Medical Association* **103**(10) 555-6.

Sharma NL, Mahajan V, Sharma RC and Sharma A (2002). Subcutaneous pheohyphomycosis in India--a case report and review. *International Journal of Dermatology* **41** 16-20.