An intriguing 'cyst' of the ear

Gideon Ze Lin Tan ,[1] Fredrik Petersson,[1] Xinni Xu,[2] Thomas P Thamboo[1]

[1]Pathology, National University Hospital, Singapore
[2]Department of Otolaryngology–Head and Neck Surgery, National University Hospital, Singapore

Correspondence to
Dr Gideon Ze Lin Tan, Pathology, National University Hospital, Singapore 119074, Singapore; Gideon_tan@nuhs.edu.sg

Received 13 February 2020
Revised 13 May 2020
Accepted 17 May 2020
Published Online First 8 June 2020

CLINICAL QUESTION

A 35-year-old woman presented with a 2 cm left ear concha fluctuant swelling that had gradually increased in size over 2 months (figure 1). There were no lesions nor lymphadenopathy elsewhere. The initial clinical impression was an ear pseudocyst. The lesion was subsequently excised. Intraoperative findings were those of a cystic lesion with a thickened anterior wall. Fragments of the 'cyst' wall were sent for histological review. Review the high quality, interactive digital Aperio slide at http://virtualacp.com/JCPCases/jclinpath-2020-206475.R2.ndpi/ and consider your diagnosis.

FIVE DIFFERENTIAL DIAGNOSES

A. Lymphoproliferative disorder.
B. Lymphohistiocytic infiltrate, possibly infectious.
C. Metastatic melanoma.
D. Rosai-Dorfman disease (RDD).
E. Soft tissue neoplasm.

The correct answer is after the discussion.

DISCUSSION

RDD, also known as sinus histiocytosis with massive lymphadenopathy, is a rare histiocytic proliferative disorder, first described as a distinct entity by Rosai and Dorfman in 1969. While traditionally regarded as a reactive condition of unknown aetiology, recent genetic studies point towards a neoplastic aetiology in at least a proportion of cases.[1]

RDD can affect all age groups but occurs more frequently in children and in young adults.[2] Although most frequently a disease of the lymph nodes, extranodal manifestations can also occur. The most frequent extranodal sites are the central nervous system, soft tissue, skin, bone, and head and neck regions.[2–4] Cutaneous RDD of the ear is extremely rare, with only seven previously reported cases.[4 5] The rarity of the presenting location (external ear) contributes to potential diagnostic pitfalls; other authors have reported initial misdiagnoses of otic RDD as non-specific chronic inflammatory lesions.

Microscopically, RDD is characterised on low power by pale areas corresponding to histiocytes, alternating with darker areas corresponding to plasma cells and lymphocytes (figure 2A). The

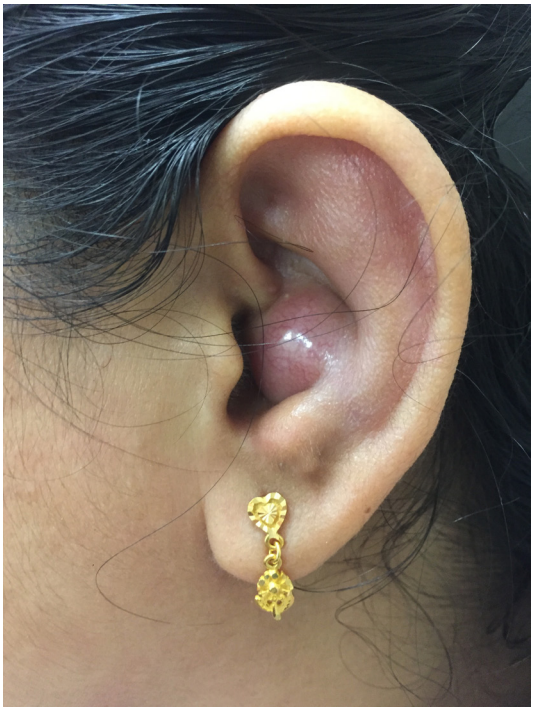

Figure 1 Clinical image of the ear swelling.

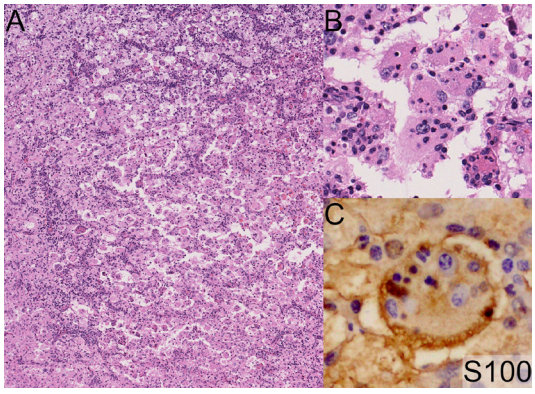

Figure 2 Scanned virtual slide (Rosai-Dorfman disease). (A) The tumour is composed of sheets of histiocytes. (B) Emperipolesis is present. (C) The histiocytes show positive staining for S100 protein.

Take home messages

- Rosai-Dorfman disease (RDD) is a rare disease characterised by a histiocytic infiltrate that stains aberrantly for the Langerhans cell marker S-100 protein. A plasma cell infiltrate and emperipolesis are characteristic.
- This case emphasises the huge spectrum of clinical presentation. RDD can present in uncommon locations without any lymphadenopathy.
- The characteristic finding of emperipolesis can be only focal, as seen in our slide. Careful search for this feature provides a helpful clue to the diagnosis, thereby averting the misdiagnosis of a non-specific lymphohistiocytic infiltrate.

To cite: Tan GZL, Petersson F, Xu X, *et al*. *J Clin Pathol* 2021;**74**:e2.

histiocytes have round, vesicular nuclei and abundant cytoplasm. Emperipolesis is characteristic although not specific (figure 2B). The histiocytes stain positively for the Langerhans cell marker S-100 protein (figure 2C) and histiocytic markers such as CD163 and CD68.[2] They are negative for other Langerhans cell markers, such as CD1a and langerin.[2]

Major differential diagnoses include inflammatory/infective conditions and other histiocytoses, such as Langerhans cell histiocytosis.

Most cases of RDD have excellent prognosis, but rare patients may show progressive disease.

CORRECT ANSWER

D. Rosai-Dorfman disease (RDD).

Handling editor Iskander Chaudhry.

Contributors GZLT and TPT conceived of the presented idea. GZLT, FP, XX and TPT all contributed towards the the writing of the manuscript. TPT supervised the project.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

ORCID iD

Gideon Ze Lin Tan http://orcid.org/0000-0002-2320-8408

REFERENCES

1 Garces S, Medeiros LJ, Patel KP, *et al*. Mutually exclusive recurrent KRAS and MAP2K1 mutations in Rosai-Dorfman disease. *Mod Pathol* 2017;30:1367–77.
2 Piris MA, Aguirregoicoa E, Montes-Moreno S, *et al*. Castleman disease and Rosai-Dorfman disease. *Semin Diagn Pathol* 2018;35:44–53.
3 Wenig BM, Abbondanzo SL, Childers EL, *et al*. Extranodal sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) of the head and neck. *Hum Pathol* 1993;24:483–92.
4 Hirt MB, Heskett J, Veerula V, *et al*. Multifocal Rosai-Dorfman disease with involvement of the Pinna. *JAAD Case Rep* 2017;3:233–5.
5 Gan L, Liu W-D, Yu W-T, *et al*. A case of cutaneous Rosai-Dorfman disease presenting with auricular enlargement as the first manifestation. *Indian J Dermatol Venereol Leprol* 2019;85:518–22.