AGAINST ALL ODDS: WHAT LIES AHEAD?
A CASE OF ROSAI-DORFMAN-DESTOMBES DISEASE

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INTRODUCTION

Rosai-Dorfman-Destombes disease (RDD) is a rare non-Langerhans cell histiocytosis (LCH) characterized by lymph node histology of sinus histiocytosis (Destombes, 1965).

Today, RDD is a rare disease of unknown etiology that may occur in any age group, but is most commonly seen in children and young adults (McAlister et al., 1990). It occurs 1:200 000 and an estimated 100 new cases per year in the United States (Mahzoni et al., 2012).

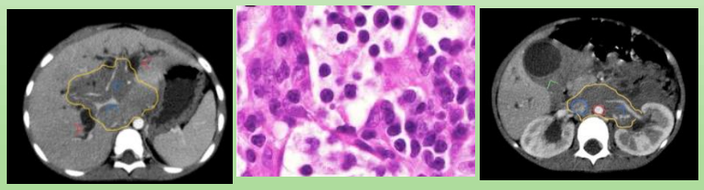
In the Philippines, there are 173 out of 4.8 million registered cases in the Philippine Pediatric Society registry under other histiocytosis syndromes. Included in the registry are sinus histiocytosis with massive lymphadenopathy, reticulohistiocytoma (giant-cell) and xanthogranuloma.

CASE SUMMARY

This paper presents a rare case of sinus histiocytosis with massive lymphadenopathy, seen in a 4yo male from Mandaluyong City, Philippines. Patient presented with a 4-week history of abdominal enlargement and jaundice. Initially assessed with Choledochal cyst; upper gastrointestinal bleeding secondary to coagulopathy. Upon further workup, the whole abdominal CT scan revealed a confluence of enlarged lymph nodes. The histopathology report of the right inguinal node revealed sinus histiocytosis, and positive immunohistochemical stains for CD68, S100 and CD1a.

This paper describes the clinical presentation, imaging, histopathology, and management of the case.

A multi-disciplinary management involving the pediatric oncologist, nephrologist, urologist, gastroenterologist, pediatric surgeon, radiologist, and pathologist was done to ensure holistic care was provided.



DISCUSSION AND CONCLUSION

RDD is a rare self-limiting non– Langerhans cell histiocytosis characterized by proliferation and accumulation of activated histiocytes within affected tissues. The etiology of RDD is not thoroughly clear and is likely not identical across the spectrum of phenotypes (Abla et al., 2018).

The diagnostic histological features of RDD include the sinus enlargement of histiocytes, possessing abundant pale or "watery-clear" cytoplasm with a large hypochromatic nucleus and prominent nucleolus. Numerous plasma cells in the medullary cords and around the venules can be seen in nodal RDD, accompanied with varying proportions of IgG4/IgG plasma cells. Emperipolesis, the trafficking of intact leukocytes through the cytoplasm, is a helpful finding but is not required for diagnosis. Immunohistochemical staining would show S100, with CD68 and variable CD163 and CD14 positivity (Abla et al., 2018).

The cornerstone of treatment has not yet been defined for RDD due to its rare occurrence (Lima et al., 2011). For patients who present with unifocal involvement, who are symptomatic, surgical resection can be done. But if unresectable, systemic therapy is the treatment recommended. For symptomatic patients with multifocal involvement, systemic therapy or palliative radiation therapy is done. Moreover, for asymptomatic patients with either unifocal or multifocal involvement, observation and surveillance are suggested. However, for any evidence of disease progression, systemic therapy is recommended.

The prognosis of RDD is correlated with the number of nodes involved. Generalized nodal dissemination and renal or pulmonary involvement suggest a poor prognosis (Ashraf et al., 2013).

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