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Case Report

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ABSTRACT

INTRODUCTION.

we report a rare case of non-Langerhans cell histiocytosis, Erdheim Chester disease (ECD). This disease is characterized by the abnormal multiplication of specific type of white blood cells called histiocytes or tissue macrophages.

CASE REPORT

A 10 years old female child presented with macrocephaly with fever and convulsion . MRI showed patchy meningitis with narrowing aqueduct treated for meningitis and vpshunt procedure. After sometimes she has recurrence of condition, hence , Biopsy of Brain was done s/o xanthomatous infiltration of tissues with histiocytes and BRAF gene mutation was positive, which was confirmative of ECD. PET–CT showed multiple bony sclerosing sites involving long bones of lower limbs, lumbosacral vertebra without any soft tissue lesion involvement. Patient has taken 3 cycles of LENALIDOMIDE (Thalidomide analogue and immunomodulator with antineoplastic properties) and Dexamethasone, with a close follow up, with a plan to repeat PET-CT but unfortunately we can't .

DISCUSSION

ECD was first described as "lipoid granulomatose" by JakobErdheim's student William Chester (1930).ECD predominantly involves the musculoskeletal system. In about half of the patients, retroperitoneum, cardiovascular system, lungs, orbits, and CNS are involved. Involvement of skin, testis, breast, and gastrointestinal system has also been infrequently reported. In ECD, involvement of lymph nodes, liver, spleen, or axial skeleton is unusual, which is common in Langerhans cell histiocytosis (LCHClinical presentation can vary from asymptomatic tissue infiltration, bony pains to multiorgan failure.

Over the last 10 years, ECD cases have been reported with increasing number as it is has became better characterized. The increase in the number of case reports would provided basis for early diagnosis, further research into the etiology and therapy of ECD. ECD being multisystem disease, multidisciplinary approach would help in better patient care and management.

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CONCLUSION

Patient was died after 15 days of therapy due to multiorgan failure .

KEYWORD:

Erdheim Chester disease (ECD)..non-Langerhans cell histiocytosis