

ALK-positive histiocytosis

ALK-positive [histiocytosis](#) was first described in [2008](#) as a systemic histiocytic disorder involving young [infants](#) and [neonates](#). Subsequently, cases of local ALK-positive histiocytosis as well as clinical presentation in adult patients have been increasingly reported in the literature.

Case reports

The current case documented the hitherto largest local ALK-positive histiocytosis lesion involving the mesentery of a 20-year-old female patient, a clinical presentation that has not been previously reported in the medical literature. Of note was the presence of numerous lymphocytes, plasma cells, and eosinophils as well as the formation of lymphoid follicles in the lesion, mimicking an inflammatory myofibroblastic tumor. Other unique histologic aspects of the current case included the nested arrangement of the histiocytes, an intravascular extension of the histiocytic proliferation into a large vein, and tumor necrosis. Notably, molecular studies revealed a novel TRIM33 (exon 12)-ALK (exon 20) gene fusion. Therefore, ALK-positive histiocytosis with TRIM33-ALK gene fusion expands the clinical, histologic, and molecular spectrum of local ALK-positive histiocytosis. Since ALK-positive histiocytosis associated with a significant inflammatory component can pose considerable diagnostic challenges, increased awareness of this particular variant of ALK-positive histiocytosis is essential to minimize the risk of misdiagnosis ¹⁾.

A case of ALK-positive histiocytosis involving the suprasellar region of a one-year-and-four-month-old boy. Through clinical, neuropathological, and genomic analyses, the patient was diagnosed with ALK-positive histiocytosis. After lesions were resected he started treatment with a combination of the three compounds vincristine, prednisolone, and crizotinib, but they did not work. Cytarabine was then added as an additional chemotherapy drug for him, and the lesions in the brain and lungs were shrunk by combining treatment of crizotinib, dexamethasone, vincristine, and cytarabine according to the RECIST (response Evaluation Criteria In Solid Tumours) ²⁾.

¹⁾
Tran TAN, Chang KTE, Kuick CH, Goh JY, Chang CC. Local ALK-Positive Histiocytosis With Unusual Morphology and Novel TRIM33-ALK Gene Fusion. *Int J Surg Pathol*. 2021 Aug;29(5):543-549. doi: 10.1177/1066896920976862. Epub 2020 Nov 27. PMID: 33243034.

²⁾
He Q, Zhang W, Li Q. Failure of crizotinib based systemic treatment in ALK positive histiocytosis involving the central nervous system: a case report and literature review. *BMC Pediatr*. 2022 May 25;22(1):308. doi: 10.1186/s12887-022-03368-1. PMID: 35614430; PMCID: PMC9131667.

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