INTRODUCTION: We wish to report a very rare case of hand – schuller – christian disease affecting both the twin male children of 6 years age. The diagnosis was made on the basis of classical clinical features, imaging findings, laboratory and histopathology findings.

Hand – schuller – Christian is a very rare disease of unknown cause. It is one of the three sub-types of Langerhans cell histiocytosis formerly called as histiocytosis x. LCH is a rare proliferative disorders of bone marrow–derived antigen-presenting cells of the dendritic cell line, also known as Langerhans cells. This disease is a spectrum of three sub-types including eosinophilic granuloma, hand-schuller-christian disease and letterer-siwe disease. We are presenting a case of 6 year old twin brothers who came with complaints of multiple painless swellings over the scalp. Subsequently they were subjected to various investigations including imaging and pathology and a final diagnosis of Hand-schuller-christian disease was given. Eosinophilic granuloma is limited to bone in patients usually 5–15 years old. Hand-Schüller-Christian disease is characterized by multifocal bone lesions and extraskeletal involvement of the reticuloendothelial system (RES) and pituitary gland, usually seen in children 1–5 years old. In Letterer-Siwe disease, there is disseminated involvement of the RES with a fulminant clinical course in children less than 2 years old. The presence of birbeck granules in the cytoplasm is characteristic. In addition tumor cells specifically express HLA – DR, S – 100 and CD1a.

CASE: 6 yrs old twin brothers presented to us with multiple, painful swellings over the scalp. On examination 1 child was severely affected with multiple painful swellings over scalp, exophthalmos, loose teeth, polyuria and polydypsia. Other child had only swellings over scalp. Skull x-ray revealed characteristic geographic skull. On CT there were multiple lytic lesions with characteristic beveled edges in b/l frontoparieto temporal bones and b/l orbital walls and mandible. MRI shows on iso to hypo T1 images and T2 these lesions are hyperintense. Serum ADH was low. Histopathology of biopsy specimen of skull lesion showed characteristic langerhans cells and birbeck granules. All these findings are consistent with Hand – schuller – Christian disease.

CONCLUSION: This is very rare case of Hand – schuller – Christian disease affecting both the twin children and our case report highlights the importance of multimodality approach including imaging and histopathology in diagnosing this disease.

CASE REPORT: Six year old twin brothers were brought to our hospital by their parents with multiple, painful swellings over the scalp. On examination one child was severely affected with multiple painful swellings over the scalp, exophthalmos, loose teeth and history of polyuria and polydypsia was given by the parents. Other child had only swellings over scalp. Skull x-ray revealed
characteristic geographic skull. On USG abdomen both the children had moderate hepatosplenomegaly. On CT there were multiple lytic lesions with characteristic bevelled edges in bilateral fronto – parieto - temporal bones and bilateral orbital walls and mandible. MRI revealed multiple, well – defined, oval lesions within the calvarial bones which are isointense to cerebral parenchyma on T1WI and hyperintense on T2WI. Serum ADH was low. All these findings are consistent with Hand – schuller – Christian disease.

BIOPSY REPORT:
- **Nature of specimen:** Biopsy from scalp swelling.
- **Microscopic Description:** Section studied shows fibro-collagenous tissue showing sheets of neoplastic Langerhans cells which have moderate amount of eosinophilic cytoplasm and nucleus with coffee – bean grooving. Langerhans cells are admixed with eosinophils, histiocytes and lymphocytes.
- **Immunohistochemistry:** CD 1a is positive within the cleaved nuclei.
- **Impression:** Langerhans cell histiocytosis.

DISCUSSION: Hand – schuller – Christian is a very rare disease of unknown cause. It is one of the three sub – types of Langerhans cell histiocytosis formerly called as Histiocytosis X.[1] LCH is a disease entity composed three rare proliferative disorders of bone marrow – derived antigen – presenting cells of the dendritic cell line, also known as Langerhans cells. Eosinophilic granuloma is limited to bone in patients 5–15 years old. Hand–schuller–Christian disease is characterised by multifocal bone lesions and extra – skeletal involvement of the reticulo – endothelial system and pituitary gland, usually seen in children 1 – 5 years old. In Letterer – siwe disease, there is disseminated involvement of the reticuloendothelial system with a fulminant course in children less than 2 years old. The presence of birbeck granules in the cytoplasm is characteristic.[2] In addition tumor cells specifically express HLA – DR, S – 100 and CD1a.

Hand Schuller Christian disease is a rare entity comprising of exophthalmos, diabetes insipidus and geographical map skull. However, the first case described by Hand[3] in 1893 had bronzed skin, hepatosplenomegaly and poor development, besides exophthalmos and geographic map skull. Schuller[4] (1915) and Christians,[5] (1920) had called in attention to this triad. It was Rowland[6] (1929) who gave histological description of this lesion in different organs while Green and Farber[7] (1942) demonstrated that eosinophilic granuloma of bone, Hand Schuller Christian disease and Letterer Siwe Syndrome have the same basic pathology. However, Lichtenstein coined the word "Histiocytosis X" to refer to full disease. Though the exophthalmos, diabetes insipidus and geographic map skull remain important for diagnosis, number of other features have also been described.

It may involve any bone, but preferred sites are the skull, mandible, spine and long bones. The lesions may be well defined or poorly defined with or without associated sclerotic borders. A variable amount of periosteal reaction can be present, ranging from a thick solid type to lamellated.[8]

In the skull, lesions usually have sharply defined borders with uneven involvement of the inner and outer table (bevelled edge sign, hole within hole). At the centre of the lytic process a button sequestrum may appear. Large lesions give the appearance of "geographic skull." In the mandible and maxilla, bone destruction may produce the appearance of "floating teeth,"[9][10]
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In the spine, typically, there is lysis of affected vertebrae leading to progressive collapse (vertebra plana, silver dollar vertebra, coin-on-edge vertebra) with preservation of adjacent disc space, which usually allows distinction from infection.[11] Areas of destruction within the centrum may be observed before collapse. A short segment kyphosis usually accompanies thoracic vertebral involvement.[12]

In the long bones, lesions usually arise in the diaphysis or metaphysis and are centred in the medullary cavity causing endosteal scalloping and eventually cortical penetration. The lesions may also appear as permeative lesions with ill-defined borders and periosteal reaction. Bone scintigraphy is of limited usefulness. CT may be useful to define the extent of the process. On MRI examination, T1 - weighted images reveal a lesion isointense to adjacent tissue and T2 – weighted images reveal high signal areas of marrow replacement.[13][14]

CONCLUSION: This is a rare case of Hand – schuller – Christian disease and affecting both the twin male children is rarest among the rare. Our case – report highlights the importance of multi-modality approach including clinical examination, serum bio – chemistry, radiological imaging and immunohistochemistry in giving a specific diagnosis to this condition.

REFERENCES:

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HAND – SCHULLER – CHRISTIAN DISEASE PICTURES

TWIN 1:

Fig. 1: Clinical picture of the twins

Fig. 2

Fig. 3: Picture showing proptosis and swellings over the scalp
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Fig. 4: x-ray AP view of skull

Fig. 5: x-ray Lateral view of skull showing typical geographical skull

Fig. 6 to Fig. 11: NECT of head showing multiple lytic lesions of the skull vault bones:

Fig. 6

Fig. 7

Fig. 8

Fig. 9
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Fig. 10

Fig. 11

Fig. 12: Showing the floating tooth

Fig. 13: MRI T1WI image showing iso–hypo intense lesions in the skull vault

Fig. 14: MRI T2WI image showing the hyperintense lesions
TWIN 2:

**Fig. 15:** Swelling in the scalp of twin 2

**Fig. 16:** X-ray AP view of the skull showing lytic lesion and scalp swelling

**Fig. 17:** Lateral skull X-ray showing the lytic lesion

**Fig. 18:** NECT image showing the large lytic lesion in left parietal and occipital bones

**Fig. 19:** Bone window image

**Fig. 20:** MRI T1WI showing lesion in the left parieto-occipital region which is iso-intense to brain parenchyma
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**Fig. 21:** MRI T1WI showing the same lesion which is hyperintense in this sequence.