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Image in Medicine

Paediatrics Section

Infantile Cortical Hyperostosis: Two Cases with Varied Presentations

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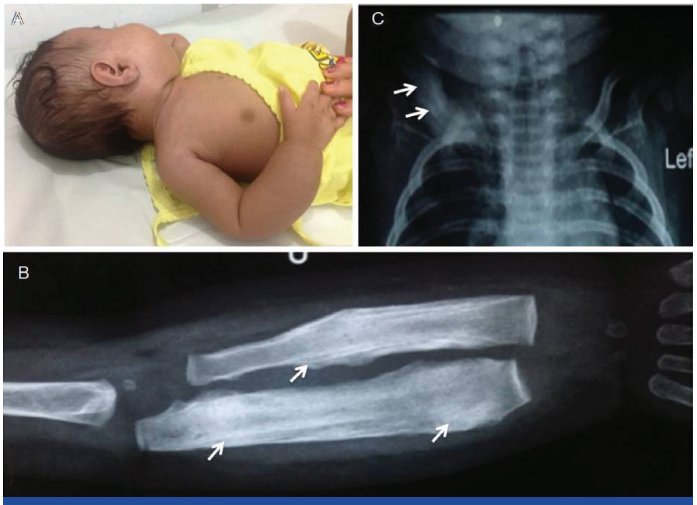
CASE PRESENTATION

Case 1: A 4-month-old female infant presented with complaints of mild grade fever, intermittent irritability and crying for last two weeks before presenting to us on 14th June 2013. There was no history of trauma, bleeding from any site, loose motion, vomiting, skin rash or any urinary complaints. She was the first child born full term to the non-consanguineous parents. Family history was not significant. Her birth and developmental history was normal. On examination, she was active otherwise and noted to have swelling over the left mandible, left forearm, and left supraciavucluar area with prominent veins. There was mild redness but no tenderness or joint swelling. There was no pallor or organomegaly. Her weight was 5.2kg (3rd to 15th centile) and length 60cm (15th centile). Caffey disease, hypervitaminosis A, scurvy, osteomyelitis, congenital syphilis, battered baby syndrome or malignancy were kept as differential diagnoses. The child was investigated as shown in [Table/Fig-1]. X ray showed thickened and irregular long bones in affected areas [Table/Fig-2a-c]. Characteristic radiological findings, laboratory investigations, and the age of onset of presentation, confirmed diagnosis of Caffey disease. A wait and watch policy was followed, and there was total improvement after three months period.

Case 2: A 2½ -month-old male infant presented with intermittent fever and irritability for last 10 day before presenting to us on 5th November 2013, that was responding to paracetamol. He was otherwise active. There was no history of trauma, bleeding from any site, loose motion, vomiting, skin rash or any urinary complaints. She was the first child born full term to the non-consanguineous parents. Family history was not significant. Her birth and developmental history was normal. His weight was 5kg (15th centile) and length 57cm (15th to 50th centile). On examination, there was only swelling of both mandibular area with some redness, and oral cavity was normal. There was no pallor or organomegaly. Caffey disease, localized osteomyelitis, battered baby syndrome, parotitis or parotid gland abscess were kept as differential diagnoses. The child was investigated as shown in [Table/Fig-1]. X-ray showed thickened irregular mandible [Table/Fig-3]. The radiological findings, laboratory investigations, and the age of onset of presentation led to a diagnosis of Caffey disease. A wait and watch policy was followed, and there was total improvement after two months period.

Laboratory parameters	Case 1	Case 2	Normal value
HEMOGRAM			
Hemoblobin	9.6 g/dl	9.8 g/dl	≥ 12 g/dl
Total leucocyte count	15,600/cumm	14,800/cumm	6,000-14,000/cumm
Total paltelet count	6.4 lac/cumm	6.1 lac/cumm	2.5-4.5 lac/cumm
ESR	110 mm/1st hr	100 mm/1st hr	Up to 20 mm/1st hr
LFT			
SGOT	54 IU/L	48 IU/L	Up to 50 IU/L
SGPT	48 IU/L	44 IU/L	Up to 50 IU/L
ALP	1320 IU/L	1240 IU/L	<800 IU/L
S.Calcium	9.6 mg/dl	10.2 mg/dl	9-10 mg/dl
S. Phosphorus	4.2 mg/dl	4.1 mg/dl	3.5-4.5 mg/dl
CRP	1.6 mg/dl	1.4 mg/dl	0-1 mg/dl
Blood Ascorbate	0.6 mg/dl	0.7 mg/dl	0.6-1 mg/dl
VDRL test	Negative	Negative	Negative

[Table/Fig-1]: Laboratory parameters

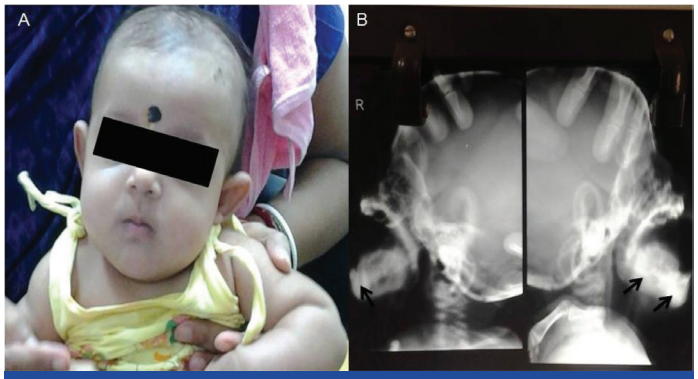


[Table/Fig-2]: A, right sided swelling of mandible, clavicle and forearm; B, X-ray showing thickened and irregular long bones in right forearm bones (arrow-mark); Panel C, X-ray showing thickened and irregular right clavicle (arrow-mark)

DISSCUSSION

First reported by Caffey and Silverman in 1945, the exact aetiology of Caffey disease or Infantile cortical hyperostosis is still unknown [1]. Two forms of the disease have been described, a classical mild infantile form (ICH) and a severe fatal form with prenatal onset [1]. The classic form most commonly presents in infants within first six months of life. The manifestations include irritability, pain, soft tissue swelling affecting one or several body parts, hyperasthesia, anorexia. Fever be present at early stages. The pain can be severe resulting in pseudo paralysis, and may cause confusion with Scurvey. Mandible is the most commonly involved site followed by scapula, clavicle, ribs and long bones. Laboratory findings include elevated ESR, alkaline phosphatase, total leucocyte count, immunoglobulin levels, CRP, platelet count, and decrased hemoglobin. Radiography is the most valuable diagnostic study. There may be florid periosteal new bone formation with pronounced cortical thickening that invlolves diaphyses sparing metaphyses and epiphyses [2].

Both our cases had the classical features described above, but in a varied manner. Though the case 1 had more of the classical



[Table/Fig-3]: A-bilateral mandibular swelling; B- X-ray showing thinkened & irregular mandible on both sides (arrow-mark).

features, case 2 simply presented with isolated mandibular involvement. When we did a literature review, we found isolated mandibular involvement to be a rare entity [2]. There is high chance of misdiagnosis or delayed diagnosis in the later case because of rarity of such presentation. It is always advisable to corroborate the clinical finding with laboratory findings in order to reach the specific diagnosis. In both our cases, the laboratory parameters were highly suggestive of Caffey disease.

Management of Caffey disease is essentially palliative, aimed at pain relief. Various treatment modalities like high dose immunoglubulins, corticosteroids, and NSAIDs (ibuprofen, indomethacin) have been tried in severe cases with varible success [3-5]. We adopted a wait and watch policy and obtained satisfactory result indicating the self limiting nature of the disease.

CONCLUSION

Caffey disease is a self-limited condition as shown by our cases. In both of the cases, a high index of suspicioun and supportive laboratory findings led to early recognition and appropriate treatment. Our case report also emphasizes the varied clinical presentation of the disease.

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