A CASE OF INFANTILE CORTICAL HYPEROSTOSIS AFFECTING ONLY THE CLAVICLE AND SCAPULA

JON WALLEY, AMERSHAM, ENGLAND

Infantile cortical hyperostosis was the name suggested by Caffey and Silverman (1945) when they published the cases of four children with deposition of subperiosteal new bone on the long bones and mandible, though, as Fairbank (1951) pointed out, Roske published the first case in 1930. Reported cases are summarised in Table I.

CASE REPORT

Boy aged two months at onset. Normal birth after normal pregnancy. Bottle-fed because the mother suffered from tuberculosis. The mother noticed a tender swelling over the child's right clavicle and that he was averse to using the right arm. Admitted with temperature of 100 degrees, pulse 120; acute osteomyelitis was suspected.

Examination—The skin overlying the clavicle was pink and slightly warm. There was diffuse tender thickening of the clavicle. No other bones were affected clinically or radiographically. Radiography showed a casing of new periosteal bone over the entire length of the right clavicle (Fig. 1). Pseudoparesis of the right arm was present.

Pathological investigations—Blood cultures were negative; blood sedimentation rate was 63 millimetres in one hour, and the Mantoux, Wasserman and Kahn tests were negative.

FIG. 1

Age two months. New bone formation encasing right clavicle.

Haemoglobin was 59 per cent (Haldane), red blood corpuscles were 3,750,000 and white blood corpuscles 11,600; differential count showed polymorphs 42 per cent, lymphocytes 48 per cent, eosinophils 2 per cent, mononuclears 8 per cent; the red corpuscles showed marked anisocytosis, moderate hypochromia and slight microcystosis.

Progress—Intramuscular penicillin was given for three weeks. At that time there was no evidence of involvement of the right scapula. Two weeks later the child seemed to be gaining weight normally and had a normal appetite. The clavicle was not further altered (Figs. 2 and 3), but clinically a small nodule was palpable in the body of the right scapula. After a further two weeks the child was beginning to use the right arm. The clavicle felt smaller. The right scapula was much increased in size, and marked thickening of the bone was shown radiographically (Fig. 4).

Five months after the onset the right clavicle and scapula were virtually the same size as the left. The child was using the arm fully and was normal and healthy with weight increasing at normal rate. A month later there was clinically no distinction between right and left scapulae and clavicles, and no other abnormalities were detected. Radiographs showed almost complete subsidence of bony changes (Fig. 6).

FIG. 2

Age two and a half months. Note marked thickening of right clavicle.

FIG. 3

FIG. 4

FIG. 5

Figure 3—Age three and a half months. No further new bone formation has occurred.

Figure 4—Right scapula at age four months. Figure 5—Left scapula for comparison.

FIG. 6

Age six months. Resolution of the bony changes is almost complete.

COMMENT

When first seen, this child was thought to be suffering from typical osteomyelitis, but the temperature was never high and toxic symptoms were never great. The condition was not influenced markedly by penicillin; indeed, two weeks after cessation of the penicillin, a similar process began in the contiguous scapula.

During eighteen months the bony changes slowly declined and this case appears to satisfy the canons of the syndrome as regards local swelling, pain, pyrexia, leucocytosis, radiographic changes and the benign course. The absence of involvement of the mandible both clinically and radiographically is, however, atypical.

I am most grateful to Mr G. P. Arden for his kindly criticism.

REFERENCES

- BRADLOW, P. A., and STEINBERG, S. H. (1950): Infantile Cortical Hyperostosis. *Journal of Bone and Joint Surgery*, **32-A**, 677.
- BURKE, F. G., and ROSS, S. (1948): Conference on Infantile Cortical Hyperostoses (Caffey-Smith Syndrome). *Journal of Pediatrics*, **32**, 441.
- BUSH, L. G., and MERRELL, O. E. (1952): Infantile Cortical Hyperostosis. *Journal of Pediatrics*, **40**, 330.
- CAFFEY, J., and SILVERMAN, W. A. (1945): Infantile Cortical Hyperostoses. *American Journal of Roentgenology*, **54**, 1.
- DICKSON, D. D., LUCKEY, C. A., and LOGAN, N. H. (1947): Infantile Cortical Hyperostosis. *Journal of Bone and Joint Surgery*, **29**, 224.
- ELLIS, R. W. B. (1939): Vitamin-C Deficiency and Periostitis of Both Ulnae. ? Scurvy. *Proceedings of the Royal Society of Medicine*, **32**, 139.
- FAIRBANK, Sir T. (1951): *An Atlas of General Affections of the Skeleton*, p. 353. Edinburgh: E. & S. Livingstone Ltd.
- FISK, C. (1947): Infantile Cortical Hyperostoses. *Journal of the Iowa State Medical Society*, **37**, 529.
- GIPSON, A. C., and CLARK, B. P. (1948): Infantile Cortical Hyperostoses. *Pediatrics*, **1**, 221.
- HALLOCK, H. (1951): Infantile Cortical Hyperostoses with a Report of Two Cases. *New York State Journal of Medicine*, **51**, 2,737.
- MACGREGOR, M., and DAVIES, R. (1949): Infantile Cortical Hyperostoses. *Lancet*, **ii**, 1,176.
- MATHESON, W. J., and MARKHAM, M. (1952): Infantile Cortical Hyperostosis. *British Medical Journal*, **i**, 742.
- MOSSBERGER, J. I. (1950): Infantile Cortical Hyperostosis. *American Journal of Diseases of Children*, **80**, 610.
- O'REILLY, J. N. (1949): Infantile Cortical Hyperostosis. *Archives of Disease in Childhood*, **24**, 67.
- ROSENBLUM, J., and GREENBERG, B. (1951): Infantile Cortical Hyperostosis (Caffey-Smyth Syndrome). *American Journal of Diseases of Children*, **82**, 711.
- ROSKE, G. (1930): Eine eigenartige Knochenerkrankung im Säuglingsalter. *Monatsschrift für Kinderheilkunde*, **47**, 385.
- SAKULA, J. (1950): Cortical Hyperostosis. *Proceedings of the Royal Society of Medicine (Section of Pediatrics)*, **43**, 242.
- SHUMAN, H. H. (1948): Infantile Cortical Hyperostosis. *Journal of Pediatrics*, **32**, 195.
- SMITHAM, J. H., and PALMER, P. (1950): A Case of Infantile Cortical Hyperostosis. *British Journal of Radiology*, **N.S. 23**, 162.
- SMYTH, F. S., POTTER, A., and SILVERMAN, W. (1946): Periosteal Reaction, Fever and Irritability in Young Infants. *American Journal of Diseases of Children*, **71**, 333.
- THOMAN, W. S., and MURPHY, R. E. (1950): Infantile Cortical Hyperostoses. *Radiology*, **54**, 735.
- WHIPPLE, R. K. (1947): Infantile Cortical Hyperostoses. *New England Journal of Medicine*, **236**, 239.
- ZEBEN, W. VAN (1948): Infantile Cortical Hyperostoses. *Acta Paediatrica*, **35**, 10.

APPENDIX
SUMMARY OF REPORTED CASES OF INFANTILE CORTICAL HYPEROSTOSIS

Author	Age at onset (months)	Presenting symptoms	Bones affected	Soft tissue swelling	Fever	Leucocytosis	Anaemia	Other features	Duration
Roske (1930)	1½	Irritability, tenderness legs	Tibiae, humerus, radii, mandible	Lower legs, arm, jaw	—	+	—	Biopsy	Clinically and radiologically normal at 9 months except some tibial bowing
Ellis (1939)	4	Fretfulness, swelling left forearm, poor weight gain	Left radius, ulnae	Left forearm	—	—	—	Suggestion of vitamin C deficiency from urine tests	Subsiding after 2 months
Caffey and Silverman (1945)	1½	Fever, irritability, tenderness left leg	Mandible, clavicles, ribs, right humerus, femora, tibiae	Both legs	+	+	—	Pleural exudate, spleen palpable	Inactive at 4 months, normal at 6 years
	2	Swelling face, fever	Mandible, ribs, humeri, radii, ulnae, femora, tibia	Face, forearms	+	—	+	Pleural exudate	Radiological changes after 10 months, normal at 5 years
	2½	Swelling face, irritability	Mandible, clavicles, left scapula, ribs, left humerus, femora, tibiae	Face, scapular region	+	+	+	Pleural thickening, seborrhoeic dermatitis	Practically normal in 10 months
	½	Swelling scapular region, restricted shoulder movement	Mandible, clavicle, right scapula, humeri, femora	Face, scapular region	—	—	—	Elevation right diaphragm, dysphagia	Radiological changes remain at 4 years
Caffey (1946)	4	Swelling cheeks, irritability	Mandible, clavicles, right radius, ulnae	Face, neck	+	+	+		Asymptomatic after 3 weeks
	18	Swelling left leg and foot, irritability, anorexia	Clavicles, ribs, right ulna, right femur, left tibia, right fibula, metatarsals	Left leg and foot	—	—	—	Night sweating	Asymptomatic after 9 months but radiological changes remain
	1½	Swelling cheek	Mandible	Face	—	+	—		Asymptomatic after 9 months but radiological changes remain
	4	Swelling face, fever	Radius, femora, tibiae, fibulae (other bones not radiographed)	Face, right leg	+				Still febrile after 4 months. No further F.U.
	20	Fever, aching joints, refusal to walk	Left clavicle, left radius, ulnae, right femur, tibiae, metatarsals		—		—	Tenderness long bones	Asymptomatic after 2 weeks
	3	Swelling face	Parietal bones, mandible, right clavicle	Face, scalp, sterno-clavicular region	+	+	+		Still symptoms after 8 weeks

* E.S.R. raised

APPENDIX—continued

SUMMARY OF REPORTED CASES OF INFANTILE CORTICAL HYPEROSTOSIS

Author	Age at onset (months)	Presenting symptoms	Bones affected	Soft tissue swelling	Fever	Leucocytosis	Anaemia	Other features	Duration
Smyth <i>et al.</i> (1946)	3½	Irritability, tenderness face	Mandible, ribs	Face	+	+	— *	Shadow superior mediastinum	Normal after 2 years Slight residual changes after 6 months
	3	Fever, irritability, swelling face	Mandible, clavicles, ulnae, femora, tibiae	Face, forearms, legs	+	—	+		
	4½	Irritability, swelling face, gastroenteritis	Mandibles, clavicles, left humerus, radii, ulnae, femora	Face, arms, left leg	+	+	+		
	2	Upper respiratory infection with fever and irritability	Mandible, ribs	Face, left scapular region	+	+	+	" Suggestion of pleurisy " in radiograph	Much improved after 9 months
	3	Swelling face and pain	Nil	Face	+	+	+		
	21	Intermittent swelling arms, feet, scalp	Clavicle, ribs, ulna, right femur	Scalp, neck, left forearm, legs, feet		+	+		
	30	Intermittent fever, irritability	Nasal bones, radius, ulna			—	—	Malnourished	
Dickson <i>et al.</i> (1947)	2	Swelling scapular area	Mandible, right clavicle, right scapula	Scapular region		+	+		Cleared in 7 months
Whipple (1947)	3	Irritability, tender swelling face	Skull, mandible, clavicles, ribs	Face, occiput	+	+	+		
Fisk (1947)	4	Swelling cheek and forearm	Mandible, right ulna	Face, neck, clavicular region	+	—	+		Almost cleared in 6 months
Burke and Ross (1948)	3½	Swelling face	Mandible, left clavicle, ribs, left ulna	Face, left arm	+	+	+	Primary T.B. lesion in chest, spleen palpable	Cleared in 6 months
Shuman (1948)	4	Swelling face, fever	Mandible, clavicles, ribs	Face, right forearm, clavicular region	+	+	+		Almost cleared in 6 months
Gipson and Clark (1948)	2	Swelling and tenderness right forearm	Mandible, clavicles, scapula, ribs, right radius, right ulna	Face, right forearm, clavicular region	+	+			Still present after 6 months
Van Zeben (1948)	1½	Swelling left leg	Clavicles, right humerus, left tibia	Left leg	—			Parents have had syphilis	Disorder disappeared after 13 months
	2	Swollen painful legs	Mandible, humeri, radii, tibiae	Mandible, legs		—	+	Brother of previous case	Died with mastoiditis 3 weeks after admission
	3	Swollen painful legs	Humerus, radius, ulna, femur, tibiae	Legs	—	+	—	Cousin of 2 previous cases	Improving radiologically 18 months later

* E.S.R. raised

APPENDIX—*continued*
SUMMARY OF REPORTED CASES OF INFANTILE CORTICAL HYPEROSTOSIS

Author	Age at onset (months)	Presenting symptoms	Bones affected	Soft tissue swelling	Fever	Leucocytosis	Anaemia	Other features	Duration
O'Reilly (1949)	2	Swelling face, fever, diarrhoea	Mandible, ribs, long bones, pelvis	Face, limbs	+	+	+	Generalised lymphadenitis	Residual broadening some bones at 15 months
Bradlow and Steinberg (1950)	3½	Tenderness jaws	Mandible (whole)	Face	100°	16,000	7.8 gms. (W.R. —ve)	Local adenitis. Added C.L.O.	Subsided 3 months. Clear 6 months
	2½	Tenderness jaws	Mandible (right ramus)	Face, right side	101°	9,250	11.2 gms. (W.R. —ve)	Pyloric stenosis (1/12) operated upon. Added C.L.O.	Subsided 9 weeks. Clear 5 months
MacGregor and Davies (1949)	1½	Swelling left thigh	Left femur, slightly right femur, later left humerus, left ulna	Left thigh	Not stated at onset (W.R. —ve)			Added C.L.O.	8 months
Sakula (1950)	3½	Swelling, tender right forearm	Radius, ulna, mandible, clavicles, tibiae, pelvis	Forearm, jaw clavicles	+	+	+	Nil	6 months
Bush and Merrell (1952)	4	Fever, irritability, tenderness right cheek	Clavicle, mandible, ulna	Face, clavicular region	+	+	+	Corticotropin given	Cleared in 4 months
Hallock (1951)	3	Tender shoulder, lump axilla	Scapula, mandible	Scapula, jaws	99°	+	+	Biopsy	Cleared in 7 months
	4½	Larger leg, irritability, pseudoparesis	Fibula, tibia, mandible	Lower leg, face	—	+	+	(Kahn —ve)	Almost clear in 6 months
Rosenblum and Greenberg (1951)	3	Irritability, tenderness face, pseudoparesis right arm	Mandible, clavicle, scapula, rib	Face	+	+	+	(W.R. —ve)	Biopsy
	3	Irritability, salivation, tenderness face	Mandible, scapula, clavicle, ribs	Face, scapula, clavicle, ribs	—	+	+	(W.R. —ve)	Cleared in 16 months
Thoman and Murphy (1950)	3	Swelling arm and thigh	Femur, humerus, scapula	Arm and thigh	—	+	—	T.B. in family. Father treated for syphilis	Cleared in 11 months
Matheson and Markham (1952)	4	Fever, facial swelling, irritability	Mandible	Face	++	+	+	(W.R. —ve)	Death. Marasmus
	4	Irritability, facial swelling	Mandible, tibiae, femur	Mandible only				(W.R. —ve)	Still not wholly resolved in 6 months
Mossberger (1950)	5½	Tenderness and swelling arms. Ecchymoses	Humeri	Both arms	++	+	+	Biopsy ante-mortem	Died 3 months
Smitham and Palmer (1950)	2	Swelling left leg	Scapula, clavicle, humeri, femora, tibiae	Lower leg	+	?	?	(W.R. —ve)	No systemic disturbances
									Slow resolution over 2 years