News, Notes and Queries

MULTIPLE BASAL CELL NAEVUS SYNDROME IN ANCIENT EGYPT

MATERIAL

TWO EGYPTIAN skeletons of the Dynastic Period are described: each shows anomalies compatible with the diagnosis of the basal cell naevus syndrome. They are in the Egyptian osteological collection of the Istituto di Antropologia, University of Turin (Cat. Nos. E225 and E 235), where one of us (M.I.S.) was carrying out research on an Italian Government scholarship in 1967.

A curious and perhaps somewhat mystical coincidence is noted.

THE SYNDROME

Multiple basal cell naevus syndrome has been adequately described only within the past decade. An important account of it was given by Gorlin and Golz (1960) and a more extensive one by Gorlin *et al* (1965). Further records appeared and the condition, although rare, is evidently commoner than was at first believed. To describe it extensively is not our purpose but its salient features are as follows.

The syndrome is a genetic abnormality probably transmitted as an autosomal dominant with incomplete penetrance. Clinically it presents as multiple basal cell naevi and skin carcinomata; often hundreds of these lesions cover the face and trunk.

Cysts, whether associated or not with a tooth, may occur in the jaws. They may be simple, occurring close to the apex of the tooth; or dentigerous and contain a tooth; or they may be primordial. These cysts range from a few millimetres in size to large lesions that may destroy much of the jaw. When infected they may produce sinuses draining into the mouth (Maddox *et al.*). Faulty dentition has also been reported.

Other skeletal deformities are less severe and may go unnoticed. These include: bifid ribs; ribs broadened at either end; and synostosis of ribs. Multiple vertebral anomalies, including scoliosis; deformed scapulae and clavicles; polydactyly, syndactyly and brachymetacarpalism—especially of the fourth metacarpals—also occur. Intracranial and other calcifications may develop, e.g. falx cerebri, the uterus, ovaries, etc. Brain tumours such as gliomata, asterocytomata and medulloblastomata sometimes occur. Ocular defects such as cataract may develop; also frontal and parietal bossing, hypertelorism, a broad nasal root and sella turcica deformities may occur. Other findings include eunuchoid features, high arched palate, facial asymmetry and increased cranial circumference.

Cases vary greatly in the number and extent of these abnormalities.

THE COINCIDENCE

Before M.I.S. left England he and C.W. discussed details of the research project, thereafter remaining in close touch by post. On 2 April 1967, C.W. wrote to M.I.S. at the University of Turin: 'Have you met any dentigerous cysts yet? If you do find any be sure to look for bifid ribs in the same body: it would be a splendid find if you

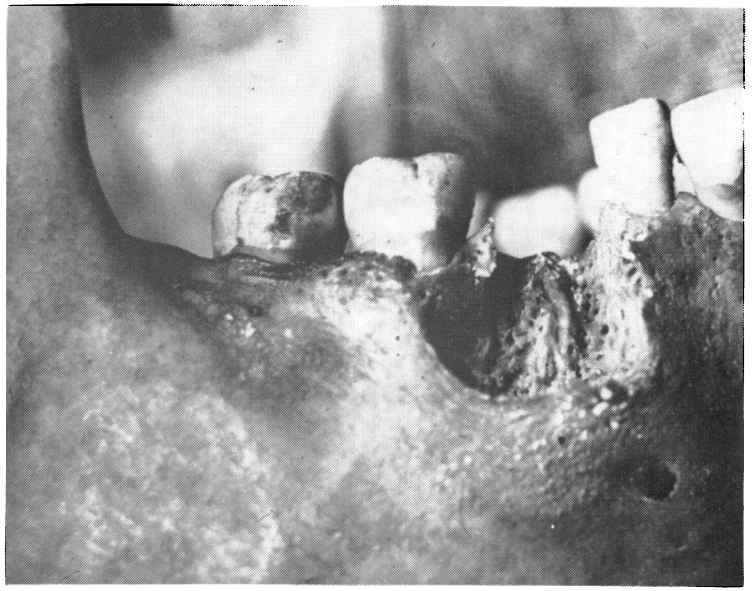

Fig. 1.

Fig. 2.

Figure 1.
(E.225) Probable dentigerous cyst cavity around shed first molar.

Figure 2.
(E.225). Bifid rib.

Figure 3.
(E.225). Incomplete fusion of sacral laminae.

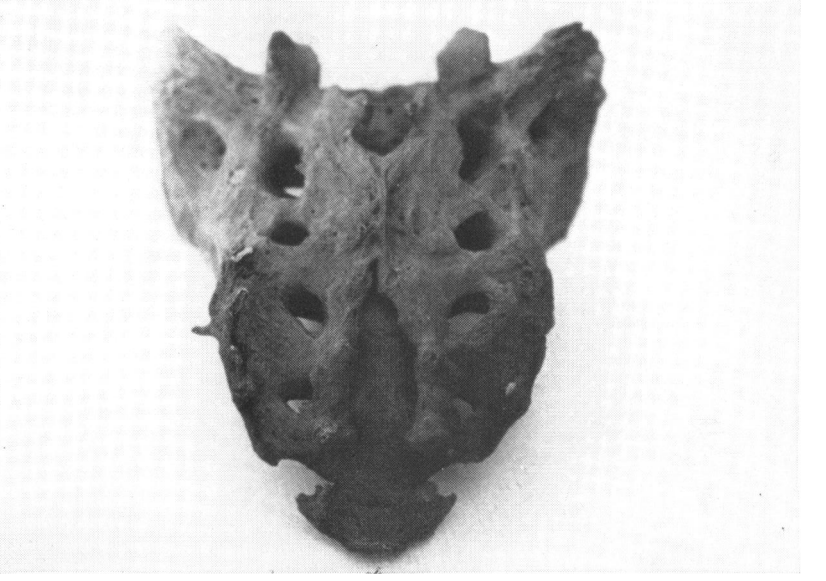

Fig. 3.

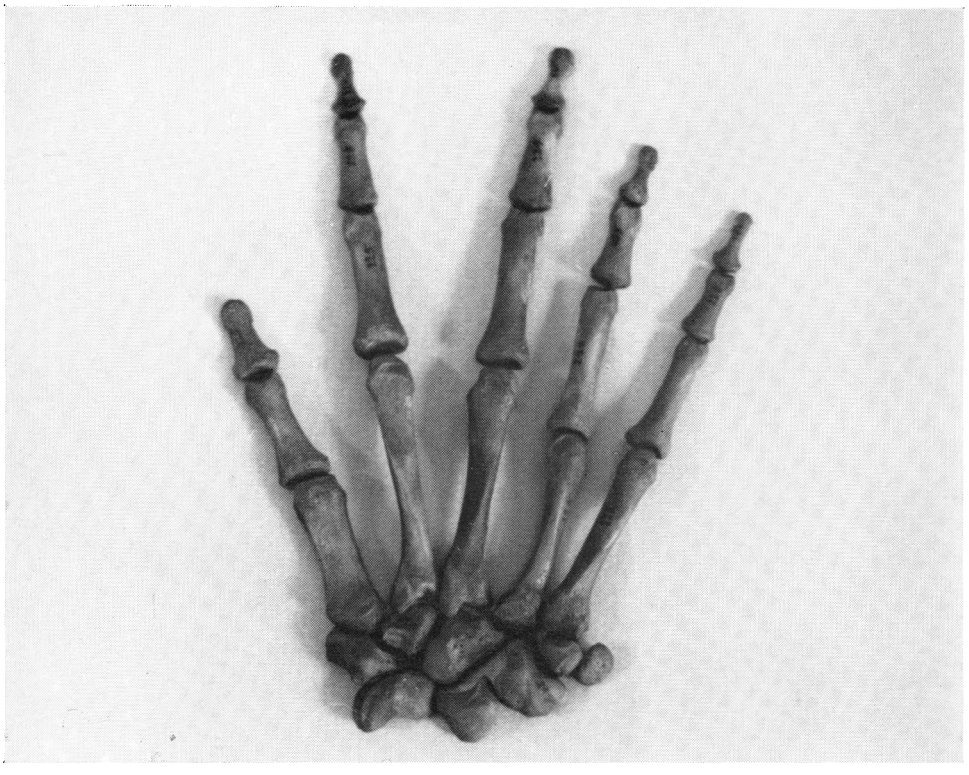

Figure 4.
(E.225). Shortening of fourth metacarpal.

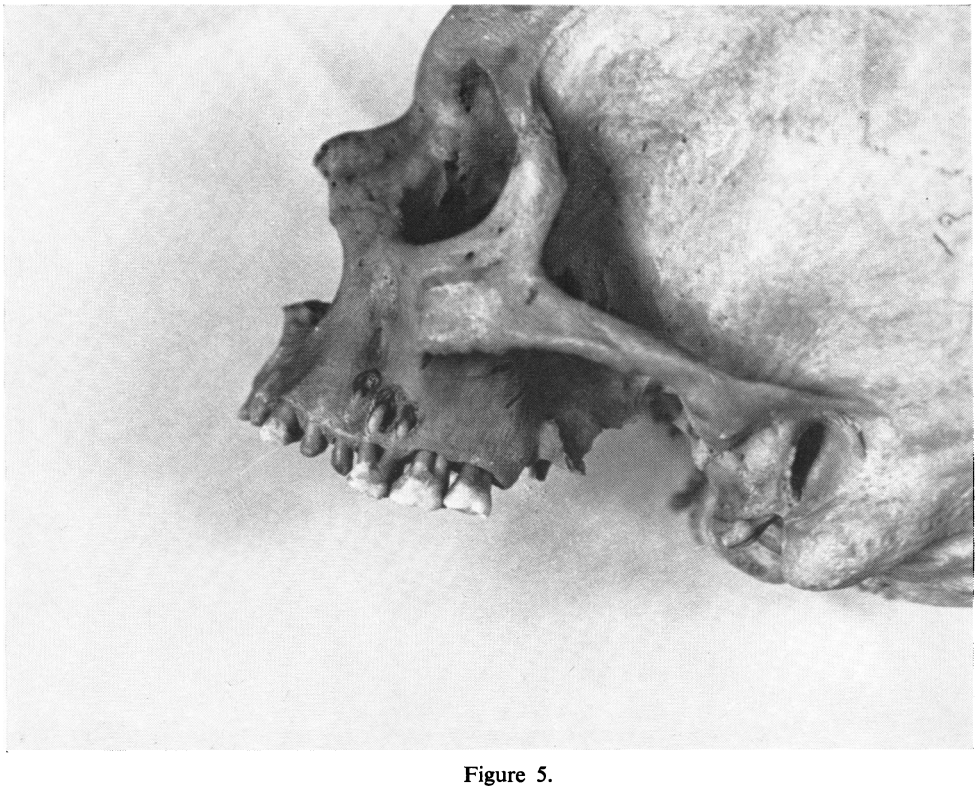

Figure 5.
(E.235). Cavitation of left maxilla.

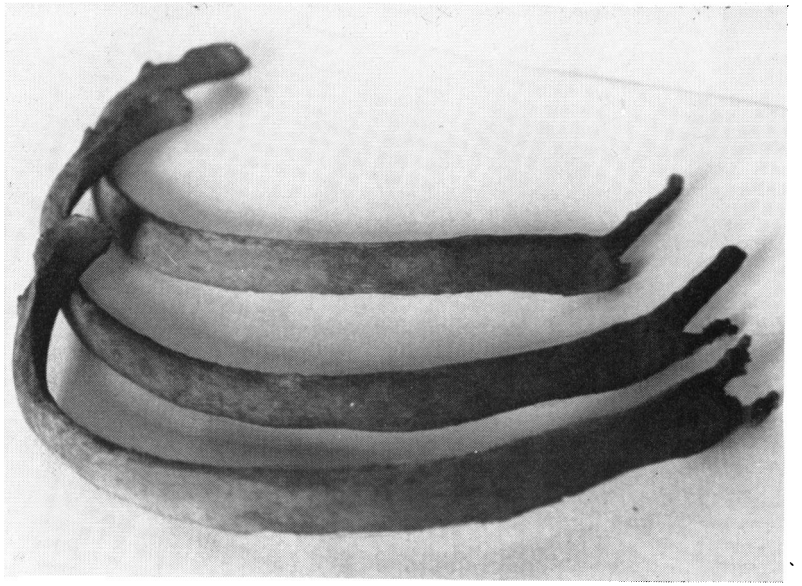

Fig. 6a.

Fig. 6b.

Figure 6a and 6b.
(E.235). Bifid ribs.

Fig. 7.

Figure 7.
(E.235). Irregularity of anterior ends of first ribs.

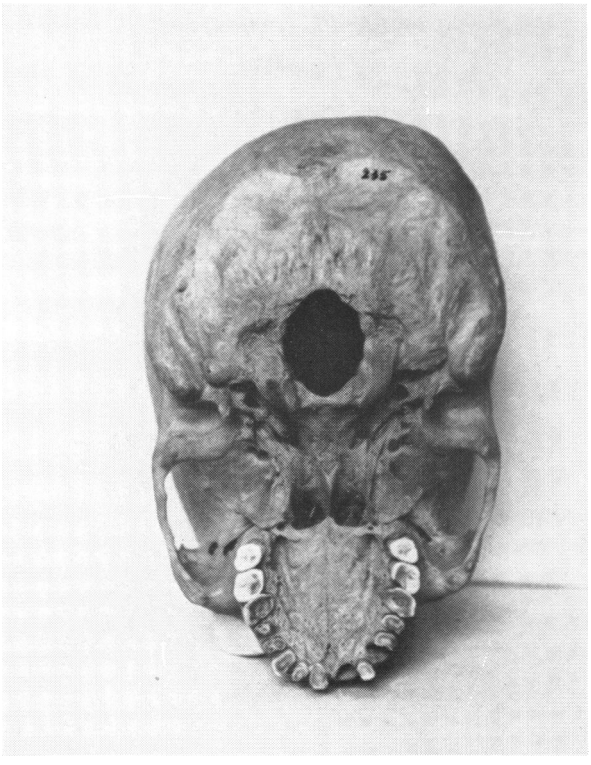

Figure 8.
(E.235). Asymmetry of occiput.

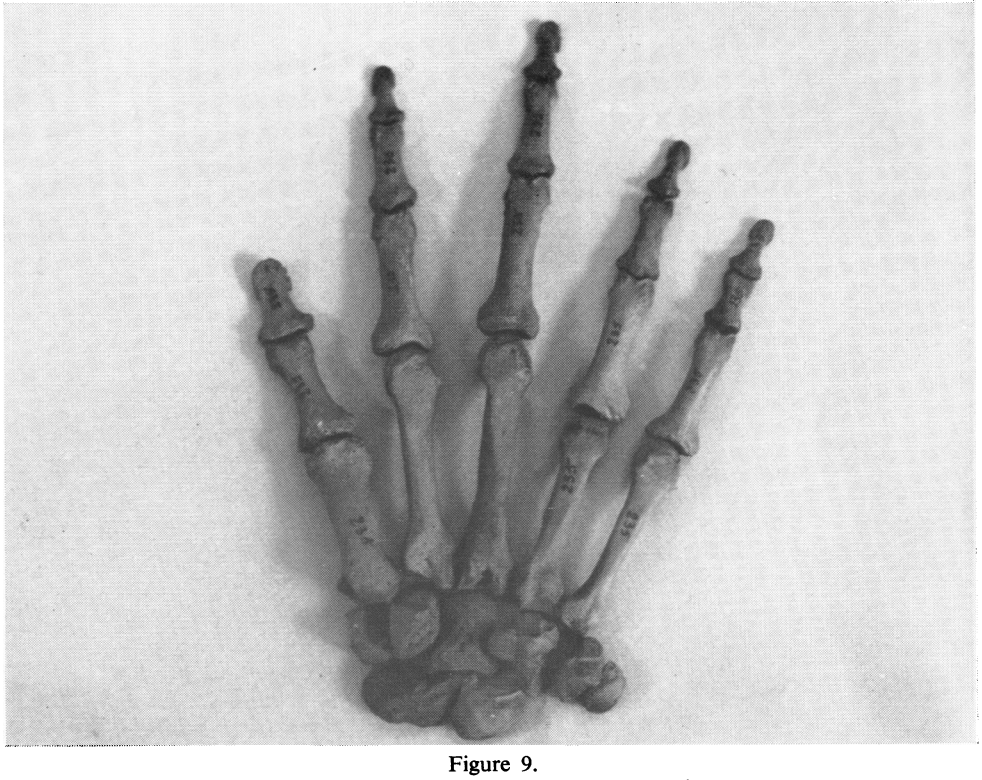

Figure 9.
(E.235). Shortening of fourth metatarsal.

discovered the dual anomaly!' On 16 April M.I.S. replied: 'you remember mentioning in your letter about the dentigerous cyst—bifid ribs syndrome, well I don't know if you believe in ancient Egyptian miracles or not but about ¾ hour before I received your letter, I was in fact examining such a case. You can imagine my excitement when I read what you wrote. It is rather like the curse of Tutankhamen in reverse!'

C. W. had only recently discussed the syndrome with Dr. E. J. Moynahan, who had seen it at Great Ormond Street Children's Hospital. C.W. then wrote back to M.I.S. giving a more detailed account of it and including further exhortations. With this new information M.I.S. re-examined the original skeleton and confirmed the finding which C.W., in little more than an optimistic pipe-dream, had suggested.

Neither of us is, in fact, addicted to beliefs in Egyptian occultism but the coincidence seems remarkable enough to deserve recording. There can be few instances in palaeopathology in which an exhortation to search for a rare disease arrived within minutes of its first discovery in ancient material.

Case 1 (E225) Male. Age 20–25 years. Excavated at Assyut.

Findings

1. Cyst in mandible near the right first molar, which had been shed during life. This was probably a dentigerous cyst (Fig. 1).
2. Bifid rib (Fig. 2). This also shows broadening posteriorly.
3. Sacrum with incomplete fusion of the laminae. Only over the dorsal aspect of the third segment is there complete development of the laminae and spine (Fig. 3).
4. Relative shortening of 4th metacarpals is present (Fig. 4).

Case 2 (E 235) Male. Age 60±10 years. Excavated at Assyut.

Findings

1. Five bony cavities are present in the maxilla. Two, situated buccally, are in relation to the mesial and distal roots of the upper right 1st molar. On the left, three are present buccally in relation to the 2nd premolar and the mesial and distal roots of the 1st molar (Fig. 5). Although the possibility of their being abscess cavities cannot definitely be excluded it seemed likely, in view of the findings in the rest of the skeleton, that they were cystic in origin. If so, they may have become secondarily infected following the pulpal exposure (Maddox *et al.*).

   There is severe tooth attrition, with pulpal exposures in the upper right 1st molar and in the left 2nd premolar and 1st molar.
2. Six bifid ribs are present—3 right and 3 left (Fig. 6 a. and b.). All are broadened anteriorly and posteriorly. A coarse, nodular irregularity at the anterior ends of the ribs, especially the first, suggests synostosis during life (Fig. 7).
3. The vertebrae show osteoarthritis and extensive ventral lipping. The reconstructed column is scoliotic but whether this scoliosis was primary or secondary is impossible to say. The sacrum shows incomplete fusion of the dorsal laminae: only over the 2nd segment is there complete development of the laminae and spine.
4. The sella turcica is slightly enlarged. The occipital bone is asymmetrical (Fig. 8) and a wormian bone is present in the right lambdoid suture.
5. Relative shortening of the 4th metarcarpals is present (Fig. 9).

Craniometric data

The following measurements were recorded on the two skulls:

Measurement	E 225	E 235
Length (gl–op)	175	189
Length (gl–in)	160	180
Parietal breadth (eu–eu)	128	137
Frontal breadth (st–st)	88	93
Frontal breadth (co–co)	107	121
Height (ba–br)	136	142
Height (projection au–br)	120	138
Orbital breadth	42	39
Orbital height	39	37
Nasal height (n–ak)	49	48
Nasal breadth	26	26
Inter-orbital breadth (mf–mf)	22	28
Nasal bones, minimum breadth	8	13
Upper facial height (n–pr)	68	65
Facial breadth (zy–zy)	116	125
Foramen magnum length (ba–o)	32	38
Foramen magnum breadth	27	28
Palate length (ol–sta)	50	49
Palate breadth (enm–enm)	33	40
Basis cranii breadth (bi–ast)	104	105
Basis cranii length (n–ba)	103	102
Facial length (ba–pr)	101	101
Mastoid height	27	28
Maximum cranial circumference	487	530

(Measurements to nearest mm.)

ACKNOWLEDGEMENTS

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MERTON I. SATINOFF
and CALVIN WELLS

SWIFT'S HALF-WAY HOUSE

ONE of the more recent developments in penology, psychiatry, and sociology has been the establishment of so-called half-way houses for ex-prisoners, narcotics addicts, alcoholics, and the mentally ill. These establishments provide a transitional environment between that of the prison, mental hospital, or sanatorium, and the everyday world to which the former inmate is returning. After confinement in institutions, apart from normal human contacts or activity, perhaps for many years, these people need the controlled environment of the half-way house to re-establish within themselves a feeling of security and self-confidence. Gradually they move, physically and then psychologically, from this protected and controlled environment to that of the world outside, wither they must eventually return to live and work. Through their stay in the half-way house, their adjustment to this world has been accelerated.

Near the end of *Gulliver's Travels*, Book IV, Gulliver spends some time in what we today would call a half-way house—the home of Captain Pedro de Mendez. In Houyhnhnm land, Gulliver has been a quasi-prisoner, quasi-servant—albeit a willing one. His frequent references to 'my master' reveal his subservience before one whom he recognizes as ethically and morally his superior. Only superior parts merit homage, he tells us. The contrast between the life of reason seen in the noble Houyhnhnms and the bestial yet manlike Yahoos unsettles him, sets up within him an irreconcilable conflict as he perceives within himself the dominance of appetite in contrast to the pure reason that he perceives in the Houyhnhnms. Forced to live in such an environment, Gulliver finds and maintains a degree of peace only by what he regards as justifiable subservience. His swoon on hearing of his fate—to have to return home—is both a measure of his attraction to a life of reason and an indication of his condition as a Yahoo himself: a Houyhnhnm would not have been moved. Almost his last words on his departure from Houyhnhnm land in Chap. X concern his own inferiority—'a creature so inferior as I.'

His original plan, to find and live upon an uninhabited island, is diverted when he