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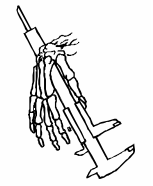

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AN INTRODUCTION TO PALAEONEUROPATHOLOGY

The examination of human remains is most often limited to the study of the skeleton, especially in Europe where mummification was not practiced and soft tissue from archaeological sites is unusual and limited to those rare instance where mummification has proceeded naturally or where adipocere has formed. Nevertheless, it is possible sometimes to infer the presence of soft tissue lesions from their effects on the skeleton; the negative impression of aneurysms of the descending aorta or the vertebral artery on the lumbar or cervical vertebrae respectively, are occasionally reported. Pathology in organ systems other than the skeleton, however, is seldom alluded to and I would like to suggest that it is possible to make some inferences about some neuropathological conditions based on skeleton changes. This is an introduction to palaeoneuropathology and is not intended to be a comprehensive review but I hope that it will stimulate interest and encourage others to pursue work in this area and to follow up some of the lines of research which I shall suggest.

It seems best to consider conditions under the conventional aetiological rubrics, infection, trauma, degenerative disease, neoplasia and congenital.

INFECTION

A number of infections may involved the central or peripheral nervous systems, including syphilis, leprosy and tuberculosis; all three give rise to changes in the skeleton that allow the diagnosis to be made relatively easily; in each case the diagnosis can potentially be confirmed by the detection of bacterial DNA using the polymerase chain reaction (PCR). All three infections may also give rise to neuropathic joints which are described below. The other infection of the nervous system most likely to be recognised in the skeleton is poliomyelitis, by inequality in the size of either the upper or lower limb bones and attended osteoporosis resulting from disuse. There have been a few descriptions of skeletons with presumed poliomyelitis and there are also depictions of the condition in paintings and statuary; you will all probably know the limestone stele from the sanctuary of the goddess Astarte at Mephis which shows the priest Ruma with an atrophied and shortened right leg, and a long stick which may have aided him in walking.

Meningitis has been suggested as the cause of death in children from medieval sites in Anatolia by Michael Schultz. His diagnosis is based on finding periosteal new bone on the inner table of the skull and is presumably considered to be a reaction to the presence of inflammatory infiltration of the dura. If the children died then it is likely that they had bacterial rather than viral meningitis which is generally a much less severe condition. At the time of writing, I do not know if the diagnosis in any of the children has been confirmed by the extraction of bacterial DNA from the skeleton; nor do I know whether the changes that Schultz has described in modern patients with the disease. It would certainly be very important to be able to demonstrate that children who die of meningitis nowadays show similar patterns of new bone formation on the skull; if they do not, then one must question the palaeopathological diagnosis and perhaps consider other causes for the lesions.

TRAUMA

Evidence of trauma is commonly encountered in human remains and in many cases it is obvious that neurological damage must have followed; this is especially the case with wounds to the head. Sometimes it is clear that the individual did not survive the trauma because there is no sign of healing but in other cases, skulls are found with extensive wounds which were caused by weapons of one sort or another and which have healed. In these cases the wounded individual lived for many months and perhaps many years, after receiving the injury and the position of the wound may provide a clue to the possibility of neurological damage. Any wound made with an edged weapon that penetrates both tables of the skull is likely to damage the underlying area of the brain and may result in paralysis if the motor area of the cortex is involved, changes in personality if the frontal lobes are damaged, visual disturbances in the case of lesions of the occipital lobes and so on. A study of modern literature relating to head injury might be useful in helping to predict the extent of neurological sequelae in past populations.

Damage to the central nervous system or the spinal cord may also be complicated by heterotopic ossification and lesions are found in up to 50% of cases. In some cases changes may be seen in the spinal cord which are very similar to those seen in DISH while in individuals with paraplegia or quadriplegia, changes may be seen in the sacriiliac joints, including periarticular osteoporosis and narrowing of the joint space. While we will not be able to recognise the latter in human remains, it may be possible to recognise the former.

One interesting cause of spinal cord compression is ossification in the posterior longitudinal ligament (PLL) in the cervical region. The condition seems to be particularly prevalent in the Japanese and is probably of relatively recent origin and it has been suggested that it has resulted from changes to a diet which is low in animal protein and high in vegetable protein in recent times. It is extremely unusual to find ossification in the PLL in European skeletons except in the presence of DISH where it may sometimes be found. It is well worth examining this site in all skeletons with DISH to see whether neurological complications may have resulted.

Spinal cord compression may also be a feature of Paget's disease and the spine of any skeleton found with this disease should be examined to determine whether or not compression of the cord was present during life.

Peripheral nerves: Many peripheral nerves are susceptible to damage, especially following fractures or dislocations to bones or joints. The most important sites of damage are shown in *Table 1*. In all skeletons with evidence of factures the possibility of neurological damage should be born in mind.

Table 1. Peripheral nerve damage consequent upon trauma

Nerve	Damage resulting from
Brachial plexus	Fracture dislocation of shoulder joint
Axillary plexus	Shoulder dislocation
Supraorbital	Injury to frontal bone
Infraorbital	Fracture of zygoma
Dental	Injury to maxilla or mandible
Suprascapular	Fracture of scapula; dislocation of shoulder joint
Radial	Fractures of humerus involving spiral groove
Median	Entrapment by ligament of Struthers attached to supracondylar spur; Fracture of elbow
Ulnar	Fracture of elbow or wrist
Anterior/posterior interosseous	Fractures of forearm
Digital nerves	Injury to fingers or toes
Sciatic	Injuries to pelvis or hip
Lateral popliteal	Fracture of head of fibula
Tibial	Fracture of tibia

DEGENERATIVE DISEASE

Of the degenerative diseases which affect the skeleton, the one which is most likely to result in neurological involvement is cervical spondylitis consequent upon alterations in the inter-vertebral disc. Intervertebral disc disease (IDD) is readily recognised in the skeleton by the presence of pitting and new bone on the end plates of the cervical vertebrae and by the presence of marginal osteophyte. When osteophytes encroach on the intervertebral foramen, then they may impinge on the emerging nerve roots and in all cases an examination of the intervertebral foramen is called for.

The intervertebral foramen: The intervertebral foramen (IVF) is formed above by the postero-lateral border of the vertebral body, the inferior border of the pedicle and anterior surface of the inferior facet joint; below it comprises the postero-late-

ral border of the uncinate process, the superior border of the pedicle and the anterior surface of the superior facet joint. The segmental nerve roots leave the spinal cord through the intervertebral foramina together with the segmental nerve and artery and in the cervical region they also traverse what is called the intertransverse space. The floor of the intertransverse space is formed by the neural groove of the vertebra below and the roof by the inferior surface of the transverse process of the vertebra above. The posterior wall is formed medially by the lateral two thirds of the anterior surface of the superior facet joint of the vertebra below and the superior border of the facet joint; laterally the posterior wall is formed by the posterior intertransverse muscles. The anterior wall is entirely muscular being formed by the anterior intertransverse muscles and the overlying longus capitis and longus colli muscles. Within the intertransverse space the cervical nerves run through the extremely narrow neural groove behind the vertebral artery. In this position the nerve may be susceptible to compression by osteophyte arising from the margins of the facet joints or by an aneurysm of the vertebral artery or a tortuosity of this vessel.

Compression of the nerves, however, is much more likely to arise from osteophyte arising from the uncinate process giving rise to neurological signs or symptoms in the arm or hand. Cervical spondylitis is common in the contemporary population; in one survey the annual incidence rates were 83.2 per 105 but approximately 203 per 105 in the group aged 50 – 54 years; the prevalence of the condition will be substantially greater than this of course, as the duration of the disease is lifelong. A radiological study of the intervertebral foramen in over a thousand patients with cervical spondylitis has shown that almost forty percent had radiologically detectable narrowing; the foramina were graded on a four point scale from 0 (normal) to 3 (most severe) with the results shown below:

Grade	N	%
0	648	59.9
1	243	22.5
2	145	13.4
3	46	4.3

The cross-sectional area of the IVF is greatly affected by disc space narrowing; with 1 mm of disc space narrowing the area is reduced by 20 – 30%; by 30 – 40% with 2 mm of narrowing, and by 35 – 45% when there is 3 mm of disc space narrowing. The anteroposterior diameter of the foramina are smaller between C4 – C6 because the uncinate processes are significantly higher at these levels. This may be the reason why the nerve roots at these levels are more susceptible to compression.

I am not aware of any comprehensive studies of the intervertebral foramen in human remains, but in some cases of IDD disease it is easy to see that the dimensions and the shape of the IVF have been altered by the presence of osteophyte and that compression of the nerve root was highly probable and the area in which neurological signs would have been experienced can be predicted from a knowledge of the dermatomes supplied by the segmental nerves.

Neuropathic joints: Both upper and lower motor neurone diseases may lead to neuroarthropathy. There are many central and peripheral causes, some of which are shown in *Table 2*.

Table 2. Some cause of neuroarthropathy

Central:	
	Syphilis
	Syringomyelis
	Multiple sclerosis
	Charcot-Marie-Tooth disease
	Trauma
	Cord compression
Peripiheral:	
	Diabetes mellitus
	Alcoholism
	Tuberculosis
	Leprosy
	Yaws
Others:	
	Chronic insensitivity to pain
	Dysautonomia

Whatever the underlying pathology, the disorganisation seen in neuropathic joints stems from sensory and proprioceptive loss which leads to relaxation of the supportive tissues around the joint which then become unstable. Normal use of the joint results in chronic injury and destruction. In the early stages, the changes affect the articular cartilage which fibrillates and fragments; subsequently the subchondral bone is destroyed, eburnation may be produced on joint surfaces and subluxion occurs. The changes may proceed very quickly and the joint may fall apart within a few weeks. In the final stages the articular ends of the bones within a neuropathic joint may be eburnated, deformed or worn away by the opposing articular surfaces; marginal osteophytes are present and may be extensive; frac-

tures are common, beginning either within or without the joint and they may show signs of repair with the formation of periosteal new bone; bone shards may be present within the joint usually with sharp, well defined edges unless secondary infection has supervened. Ankylosis of neuropathic joints is uncommon, except in the spine, and some of the debris from the joint may not be recovered during excavation.

The pathological features of the condition are similar whatever the underlying neuropathology, but the distribution of the changes seen does vary considerably and may give some indication of the primary diagnosis (see *Table 3*).

Table 3. Common sites of involvement in neuroarthropathy

Disease	Sites involved
Syphilis (tabes dorsalis)	Spine, hip, knee, ankle
Syringomyelia	Spine, shoulder, elbow, wrist
Diabetes mellitus	Tarsal, TMT, MTP
Alcoholism	MTP, IP of feet
Amyloidosis	Knee, ankle
Meningomyelocoele	Ankle, tarsal
Congenital sensory neuropathy	Knee, ankle, tarsal, MTP, IP of feet
Idiopathic	Elbow

TUMOURS AND TUMOUR-LIKE CONDITIONS

Conditions under this head include meningiomas, neuromas and neurofibromatosis, all of which may affect the skeleton.

Meningioma: Meningiomas are said to account for approximately 15 – 20% of all primary intra-cranial tumours and they are the commonest cause of pathological vascular markings on the skull. It is thought to arise from arachnoid cap cells, specialised cells within the arachnoid granulations, although it is not clear that these are the only cells that can form a meningioma. Nevertheless, meningiomas are found in those sites where the arachnoid granulations are most frequent, that is, the superior sagittal sinus, the basal cisterns, the transverse sinus and other dural sinuses.

Although they are generally benign, meningiomas are known rarely to metastasise, with the lung, abdominal viscera and the skeleton being the most common sites for distant spread.

Meningiomas are recognised in the skeleton by pressure defects in the inner table of the skull that may also erode through the outer table. They are supplied by aberrant branches of the intra-cranial vessels and they may also stimulate hyperostosis. When the tumour arises in the cranial vault, one or more branches of the middle meningeal artery may be enlarged and this may be accompanied by enlargement of the foramen spinosum. The hyperostosis produced may affect both tables of the skull and the appearance may be similar to that seen with an osteosarcoma.

There have been relatively few descriptions of meningioma in the palaeopathological literature and those that have been reported have most often been of the type in which hyperostosis has been a prominent feature; in fact, it is the new bone on the external table of the skull that generally draws attention to the lesion, although they may also be detected when the lesion erodes through the outer table. In the latter case, the lesion is brought to light by a hole in the skull. Lesions that are confined inside the skull come to light only when the skull is broken or when the interior of the skull is examined radiologically or endoscopically.

The incidence of meningioma in the modern population is said to be approximately 2/105; figures for the prevalence of the condition do not seem to have been published, but would be greater that this. The prevalence in some palaeopathological conditions, however, has been quoted as greater by two or three orders of magnitude. For example, Campillo found five cases among 3,000 skulls from different sites in Spain, giving a crude prevalence of 0.17% (95% CI 0.05 – 0.38%) while in a site from southern England, I found crude prevalences of 2.41% for males and 1.19% for females (95% CI 0.29 – 8.43% and 0 – 6.45%, respectively).

These prevalences for past populations seem extraordinarily high when compared with modern data but it has to be remembered that the modern data are based on cases that come to operation. The majority of meningiomas that have been described in human remains are small and it is likely that the individuals in whom they were found would have been unaware of them during life. It is probable that only a small fraction of meningiomas in the general population become large enough to produce serious neurological signs or symptoms and to require surgical intervention. It would be extremely interesting to determine the prevalence of meningioma from radiographs of the skull taken in patients with non-neurological conditions, for example, and to determine the prevalence in other skeletal assemblages to compare the frequency both in space and time.

Neuromas: Nerve cell tumours may affect the skeleton by causing pressure defects. In the spine, a

neuroma may enlarge the inter-vertebral foramen where they may have to be differentiated from the changes caused by vertebral aneurysm or tortuosity. An acoustic neuroma may be recognised in the skeleton by enlargement of the internal auditory meatus (IAM) and this may be confirmed by x-ray. In at least 40% of cases, tumours of the eighth nerve are bilateral and so both IAMs may be enlarged.

Neurofibromatosis: In this condition, skeletal abnormalities are prominent and it is these that are likely to alert the palaeopathologist to its presence, although the changes consequent upon the presence of multiple neuromas should also be visible. The skeletal abnormalities may include defects in the greater or lesser wings of the sphenoid, defects in the lambdoid suture – generally on the left hand side, scoliosis, bowing of the long bones sometimes accompanied by fractures and pseudarthrosis. All the skeletal changes are the result of mesodermal dysplasia.

CONGENITAL DISORDERS

Neurofibromatosis is one of the more common congenital conditions, having an incidence of approximately 1 in 3,000 births but is has been described above as fitting more easily into the section on tumours of the nervous system. The only other congenital disorder to be considered here is spina bifida.

Spina bifida: The term spina bifida is most frequently used by palaeopathologists to describe failures of fusion of the laminae of the sacrum; it may also often be referred to as 'spina bifida occulta'. The use of this term to describe a condition that is clinically benign should be abandoned and another without clinical overtones used instead; sacral malfusion might be preferable and being descriptive and neutral.

True spina bifida – dysraphism – involves failure of midline fusion of the sacrum or vertebrae and the neural tube; the lesion may also include the gastrointestinal or genitourinary systems. When the lesion is uncovered it is referred to as open; when covered by skin, then it is referred to as closed, or occult. In both cases, vertebral anomalies are generally present. The interpedicular distances of the vertebrae at the level of the abnormal neural and menigeal tissue are widened. In addition there may be vertebral hypoplasia, hemivertebra and fusion of the laminae or pedicles; all these changes should be evident in the skeleton. In patients with diastematomyelia, where the spinal cord is split in two longitudinally, there may be a bony spur between the two parts of the spinal cord, extending from the neural arch forwards, sometimes fused anteriorly to the vertebral body. Again, this anomaly should be visible on examination of the skeleton.

The incidence of spina bifida varies from country to country but is approximately 1 – 2 per thousand births. It is probable that it was higher in the past when diets may have been relatively deficient in folic acid and it seems worthwhile making a close examination of the skeletons of young children to ensure that none of the skeletal markers of spina bidifa is present.

OTHER CONDITIONS

I would like to mention one other condition that does not fit easily into the rubrics considered above, and that is raised intracranial pressure. In children, raised intracranial pressure may cause convolutional markings on the skull, giving rise to the so-called beaten copper appearance on radiographs. With so many children being represented among most skeletal assemblages, the possibility that some may have died as the result of raised intracranial pressure should be borne in mind and the internal table of the skull examined for evidence of convolutional markings; perhaps incorporating radiography in suspicious cases.

CONCLUSION

The account which we are able to give of disease in past populations is undoubtedly limited severely by the lack of soft tissue for examination. I hope that I have been able to show that, nevertheless, it should be possible to infer something about the presence of neurological disorders from the study of the skeleton and that others will be encouraged to undertake the study of palaeoneuropathology.