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PATHOGRAPHY OF LADY IMAKHETKHERRESNET, SISTER OF PRIEST IUFAA

PATOGRAFIJA IMAKHETKHERRESNET, SESTRE SVEĆENIKA IUFAAE

Eugen Strouhal¹, Alena Němečková², Fady Khattar³

Abstract

Lady Imakhetkherresnet was buried at the age of 35-45 years in the southern corridor of a well preserved shaft tomb of priest Iufaa at Abusir (end 26th Dynasty, 625 BC). The tomb was excavated by the Czech Institute of Egyptology from 1994 to 2004. Morphometric, genetic and epigenetic features linked her by blood to Iufaa; epigraphic evidence concluded that she was his sister.

Her pathography includes the usual tooth diseases, and early stage vertebral osteophytosis and degenerative osteoarthritis. She also suffered a spiral fracture of both right lower leg bones.

A large smooth-walled cavity was found in her sacrum, moulded by the pressure of a relatively hard tissue mass. Its extent and lobulated form were first assessed macroscopically and then by standard radiography. CT sections revealed wide cavities extending from the spinal canal to both 2^{nd} sacral foramina and to the left 3rd sacral body. A benign neurilemmoma was diagnosed by macroscopy and radiography, and confirmed by histology. This benign tumour is the first of its kind and localization to be identified in palaeopathology and in the history of medicine.

Keywords: pathography, bone and tooth diseases, neurilemmoma, Abusir, 26th dynasty, Egypt

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INTRODUCTION

Pathography, the influence of diseases on the life and work of famous persons, can also be described in people of antiquity. New archaeological investigations, especially in Egypt and the Middle East, have been discovering more and more historical personages of that period. They are identified by name, title and affiliation not only thanks to new finds of written sources (papyri, reliefs, or clay tablets), but also by direct finds of their bodies thanks to modern medical, natural history and technical methods. Specialists trained in *palaeopathology* are now able to discover and diagnose diseases that leave traces in their bones and teeth.

ARCHAEOLOGICAL BACKGROUND

Lady Imakhetkherresnet was unknown before a team of archaeologists of the Czech Institute of Egyptology in Prague directed by Ladislav Bareš found her body during an investigation of a great shaft tomb located half a kilometre to the south of the Pyramid of King Neferirkare at Abusir. This investigation started in 1994 (Bareš & Smoláriková 1996) and ended in the spring of 2004.

The tomb has been spared from pillage. According to texts on the sarcophagi, it belongs to priest Iufaa, who was the administrator of the King's palaces, most probably at Sais, the capital of Egypt during 26th Dynasty. His double sarcophagus was opened in 1998 and his mummy was taken out for examination. He was found to have died a premature death at the age of 25-30 years, at which time the construction of his luxurious tomb had not been completed (Fig.1, burial no.1; Bareš & Strouhal 2000, Bareš 2003, Strouhal 2003).

During the excavation season of 2001, another mummy of a 35-45 year old woman, later identified as Imakhetkherresnet, was found in a truncated end of a 26 m long corridor to the south of Iufaa's tomb (Fig.1, burial no.2). (Bareš et al. 2002).

In front of Imakhetkherresnet's double coffin there was another 55-65 year old male (Fig.1, burial no.3), presumably buried there soon after her burial. His wooden coffin was deteriorated by humidity to such an extent that the corpse could not be identified by name and titles.

Mummies of two other males, Neko and Gemenefherbak, the priests and administrators of the King's palaces, were discovered in two burial

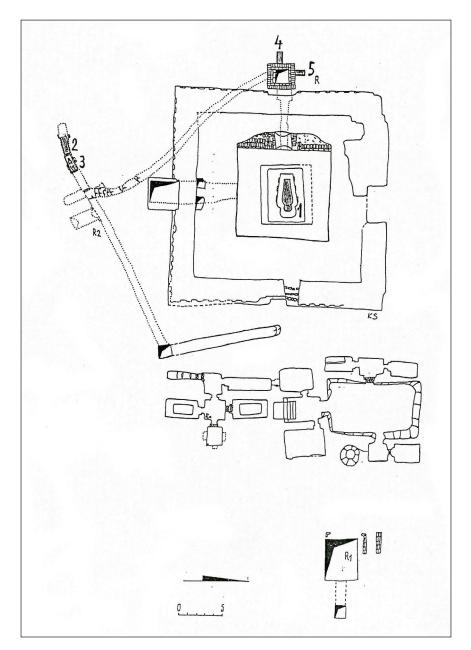


Figure 1. Schematic plan of Iufaa's Shaft Tomb at Abusir. Double coffin with mummy of Imakhetkherresnet was found in the west dead end of the southern corridor marked R2.

Slika 1. Nacrt grobnice Iufaae u Abusiru. Dvostruki lijes s mumijom Imakhetkherresnete nalazi se u zapadnom dijelu južnog koridora označenog kao R2. niches near the bottom of a small west shaft of the tomb (Fig.1, burials no. 4 and 5).

BLOOD RELATION WITH IMAKHETKHERRESNET

Several metric, epigenetic, and descriptive features as well as identical congenital anomalies confirmed a blood relation between Iufaa and Imakhetkherresnet (Strouhal 2002 a,b). Moreover, palaeographic evidence of the identical mother's name Ankhtisi confirmed that Imakhetkherresnet was Iufaa's sister. No title was inscribed on either of her coffins (Bareš et al., 2002).

Still closer similarity in the above features was found between Imakhetkherresnet and the anonymous male. The 20 year gap between them at death and short time elapsed between their burials suggest that they were daughter and father rather than siblings. They were buried at the dead end of a corridor made especially for them at the time of Iufaa's tomb construction, before his burial. The corridor was (perhaps later) connected to the 24 m deep tomb bottom by a steeply descending S-shaped passage.

DEMOGRAPHIC DATA OF IMAKHETKHERRESNET

Imakhetkherresnet's skeleton is gracile, with a relatively well-developed muscular relief and bones mineralized in accord with her age at death. Her stature, reconstructed according to tables by Trotter and Gleser (1952) for Afro-Americans, which suit better the proportions of the Ancient Egyptians than the tables for American Whites, was underaverage, only 151.5 cm.

As regards the *age at death*, her cranial sutures are in progressed fusion, including the lambdoid suture and pterion (similarly as in the skull of her brother Iufaa). Abrasion of the teeth was medium (stages 4-5), except for the upper left premolars with stage 6 abrasion (according to Brothwell 1972 modified by Strouhal & Jungwirth 1984). A stage 8 (by Todd 1920) *facies symphysialis*, mild osteophytosis of the spine, and early degenerative arthritis of the big joints suggest an age at death of 35-45 years.

PATHOGRAPHY OF IMAKHETKHERRESNET

In her young age, Imakhetkherresnet was a well-developing woman. Her two small carpal bones (left *capitatum* and *hamatum*) were fused most probably due to a trauma of the ulnar side of carpus, if not congenitally. That she was spared from heavy work is suggested by a mild (stage 2-3) osteophytosis of the spine only in the lumbar section (according to Stloukal & Vyhnánek, 1974).

Spondylarthritis of $C_{3.4}$ with early foci of eburnation speaks of frequent rotation of her cervical spine. There was only a minimal degenerative arthritis of the big joints, especially of the hips, indicating that she seldom carried or lifted weights.

Judging by caries-free teeth, her diet was well balanced but rough, as of other Egyptians of her age, which led to progressed tooth abrasion and medium stage retraction of the alveolar process (according to Brothwell 1972). She lost three lower teeth: the right P_2 and the left lower M_1 and M_3 .



Figure 2 Spiral fracture of the right crural bones of Imakhetkherresnet from anterior side. Slika 2. Spiralna fraktura kostiju desne

potkoljenice Imakhetkherresnete. Pogled sprijeda. Judging by her medium deep sulcus praeuricularis she had given birth to one or more children. There is no information about her husband.

In her young adult age she fell and sustained a spiral fracture of the right lower leg bones between the middle and distal third of their diaphysis (Fig. 2). It was an open fracture complicated by osteomyelitis, as suggested by three openings on the bone surface. Nevertheless, the X-ray revealed perfect healing of the medullar cavity without remnants of fracture lines or abscess cavities. A relatively thin callus, minimal dislocation of the bones and the synostosis of the proximal tibiofibular joint prove than the fracture was treated by immobilisation and most probably by medical intervention (Strouhal et al., 2003).

The most unusual pathology was a long lasting benign tumour in her sacrum (Strouhal & Němečková 2004).

THE TUMOUR OF IMAKHETKHERRESNET

The posterior view of her six-piece sacrum (by sacralization of the fifth lumbar vertebra) reveals the presence of a large, well rounded and slightly oval orifice (max. diam. 25 mm) in the 2nd sacral segment right of the crista sacralis mediana. It merges with the right 2nd sacral foramen and leads into a smooth walled circular and A-P flat cavity inside the bone excavated by the pressure of a relatively hard growing tissue mass. An irregularly edged small opening to the left of the crista sacralis mediana of the same sacral segment resulted from a secondary fracture of the compact bone, thinned by the steady pressure of that same tissue growing inside the bone (Fig. 3).

In the anterior view, a secondary perforation is also visible in the middle of the 3^{rd} sacral segment. The anterior wall in the middle of the 3^{rd} sacral segment is thinned and bulges anteriorly for about 2 mm. It has been perforated secondarily by the expanding growth, as evidenced by the irregular edges of the opening (Fig. 4).



Figure 3. The sacrum in posterior view showing the large orifice in the right half of the second segment and a small secondary perforation in its left half as well as several similar perforations in the middle of the third sacral segment.

Slika 3. Stražnja strana krstače pokazuje širok otvor u desnoj polovici drugog segmenta i mali sekundarni otvor u njegovoj lijevoj polovici. Vidljivo je nekoliko sekundarnih otvora u sredini trećeg krstačnog segmenta.



Figure 4. The six-piece sacrum in anterior view with bulged anterior wall with a secondary opening in the middle of the third segment.

Slika 4. Šestodijelna krstača s izbočenjem prednjeg zida sa sekundarnim otvorom u sredini trećeg segmenta. Pogled sprijeda.

STANDARD RADIOGRAPHY OF THE TUMOUR

In the postero-anterior view, a large circular cavity (28mm in diameter) on the right side of the 2^{nd} sacral body is delineated by a smooth edge of increased density and merges with the 2^{nd} right sacral foramen (Fig. 5).

Two smaller cavities project to the left of the crista sacralis mediana of the same sacral body. The lower is vertically oval and the upper is semicircular. They are separated by a layer of dense trabeculae. However, CT sections show a single common cavity, merging with the 2nd left sacral foramen (see further).

The largest circular cavity (30 mm in diameter) in the 3^{rd} sacral body is located mainly to the left, but a third of it goes beyond the crista sacralis mediana, extending to the right. Its dense margin is interrupted by a fusion with the 3^{rd} left sacral foramen.

In the slightly oblique lateral projection, the described cavities form a tuft of three partially superimposed lobes at the level of the 2^{nd} and 3^{rd} sacral bodies (Fig. 6).

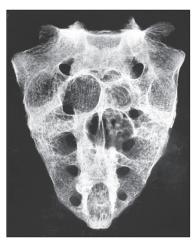


Figure 5. Standard postero-anterior radiograph of the sacrum with two big and two small well delimited cavities.

Slika 5. Standardni P-A radiograf krstače na kojem su vidljive dvije male i dvije velike dobro ograničene šupljine.



Figure 6. Standard lateral radiograph of the sacrum showing a tuff of three superimposed cavities.

Slika 6. Standardni postranični radiograf krstače na kojem su vidljive tri superponirane šupljine.

CT IMAGING OF THE TUMOUR

In order to ascertain in detail the number and location of the lobes, the sacrum was scanned using a Siemens Somatom Plus 4 Spiral Computed Tomography Scanner with the following parameters: slice thickness 3 mm, table movement per rotation 4.5 mm (pitch 1.5), reconstruction index 1 mm.

The scanned images were transferred to a Silicon Graphics O2 workstation with Virtuoso (Siemens) 3D manipulation software to produce coronal (3mm index), sagittal (3.56 mm index) and axial reconstruction (1 mm index).

Coronal (frontal) sections from posterior view (Fig. 7), show a definite widening of the sacral canal from which abnormal cavities with arched outlines and sclerotic edges project.

The first cavity in the right half of the 2^{nd} sacral body ($28 \times 28 \times 24$ mm) fused with the 2^{nd} right sacral foramen and caused a large defect in the dorsal aspect of the sacrum.

The second cavity in the left half of the same body was smaller $(13 \times 14 \times 22 \text{ mm})$. It fused with the 2nd left sacral foramen and was related to the above mentioned small *post mortem* defect in the thinned left dorsal aspect of the sacrum.

The third cavity in the left half of the 3^{rd} sacral body extends across the midline into the medial part of the right half of the same sacral body, reaching there its biggest dimensions ($30 \times 26 \times 22$ mm). It fused with the smallest consistent cavity no. 2 left in the 2^{nd} sacral body and with the 3^{rd} left sacral foramen causing the above mentioned bulging and perforation of the thinned compact bone. The described cavities were created by the pressure of the growing lobes of a single benign tumour, as demonstrated by their mutual connections.

Axial section at the level of the second sacral body shows a transversal outline of the cavities – the left one oval, the right one oblong – and their posterior connection as well as their relation to the widened sacral canal and to the opening on right side (Fig. 8).

The section through the third sacral body shows a regular oval outline of the largest and thickest third cavity, bulging antero-posteriorly (Fig. 9).

In a *sagittal section* from the right side the bulging anterior wall of the third sacral segment caused by extension of the largest cavity no. 3 can be observed (Fig. 10).

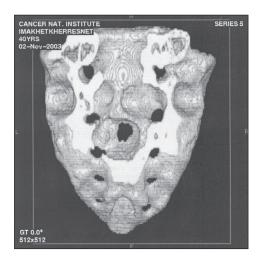


Figure 7. A coronal section from posterior side shows the maximum extent of the two biggest cavities, their fusion with the sacral openings and the secondary perforation in the middle of the third segment.

Slika 7. Koronalni CT rez straga pokazuje najširi promjer dviju najvećih šupljina, njihovo spajanje s otvorima krstače i sekundarnu perforaciju u sredini trećeg segmenta.



Figure 8. An axial section from the top across the second sacral segment shows the limited anterior reach of both cavities, their different sizes and outlines and their relation to the widened sacral channel. There is only one common cavity in the left half of the vertebral body.

Slika 8. Aksijalni presjek drugog krstačnog segmenta, pogled odozgo. Vidljivi su ograničeni prednji dosezi obje šupljine, razlike u njihovoj veličini i njihov odnos prema proširenom krstačnom kanalu. Postoji samo jedna šupljina u lijevoj polovici.

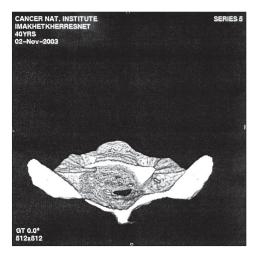


Figure 9. An axial section from the top across the third sacral segment demonstrates the oval shape and large extent comprising almost the whole vertebral body.

Slika 9. Aksijalni presjek trećeg krstačnog segmenta, pogled odozgo. Vidljiv je ovalan oblik šupljine koja svojom veličinom zauzima gotovo cijelo tijelo kralješka.



Figure 10. In a sagittal section from the right side the bulge of the anterior wall of the third sacral segment by pressure of the growing tumour broadly adhering to the sacral canal can be distinguished.

Slika 10. Sagitalni presjek, pogled s desne strane. Izbočina prednjeg zida trećeg krstačnog segmenta posljedica je pritiska tumora čija šupljina široko prianja na krstačni kanal.

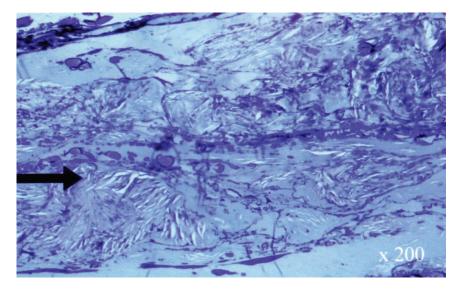


Figure 11. Histological section of an area of the tumour with the Antoni A structure (the arrow points to spindle shaped Schwann cells).

Slika 11. Histološki presjek tumora sa strukturom Antoni A (strelica pokazuje vretenaste Schwanove stanice).

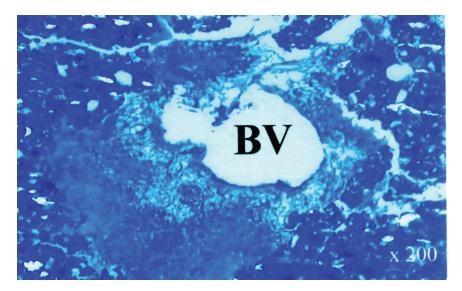


Figure 12. Histological section of an area of the tumour with the Antoni B structure (BV = blood vessel).

Slika 12. Histološki presjek tumora sa strukturom Antoni B (BV=krvna žila)

MORPHOLOGICAL DIAGNOSIS OF THE TUMOUR

To diagnose/identify pathological cavities inside Lady Imakhetkherresnet's sacrum, we had to consider different benign and malignant tumours and tumour-like infection changes (see Strouhal et al., 2003; Strouhal & Němečková, 2004). A single cavity with three lobes was identified. All originated from a solid benign tumour growing slowly from the sacral nervous plexus. The host reacted by forming a well-defined and smooth sclerotic bone capsule.

In this specific case there are two kinds of benign tumours derived from the nerve myelin sheath that may have caused the cavity: neurofibroma and neurilemmoma (schwannoma, neurinoma; Aufderheide & Rodriguez – Martín, 1998: 385).

The macroscopic morphology of the latter as described by Škorpil (1950: 558) suits best the appearance of Imakhetkherresnet's hollow in the sacrum. According to Škorpil, neurilemmoma is "a well delimited tumour of globular or egg shape, nut to apple in size, rarely of larger dimensions. Its surface is knotty or lobulated. In some localisations, especially in the spinal canal, it can be of hourglass shape by growth of part of the tumorous matter through the foramen intervertebrale, while the other part remains deposited intradurally".

In this case the hourglass shape of the tumour can be attributed to the penetration of the tumourous mass through both the 2^{nd} and the 3^{rd} left sacral foramina, and through the large orifice in the right posterior wall of the 2^{nd} sacral segment. The extent of the extrasacral expansion of the tumour is uncertain, but it could have become symptomatic by pressure on the sacral plexus and/or by bulging under the skin of the back.

HISTOLOGICAL DIAGNOSIS

Morphological diagnosis could be confirmed thanks to two tiny remnants of the soft tissue preserved at the edge of the large orifice in the second sacral body. Light microscopy showed two characteristic structures of neurilemmoma (Figs. 11-12, Strouhal & Němečková, 2004).

Of the four immunohistochemical tests for neurilemmoma (Kleinhus & Cavenee, 1999), the test for the presence of S-100 protein and the test for Leu-7 (CD 57) yielded negative results. The other two tests for glial fibrilar acidic protein (GFAP) and for the epithelial membrane antigen (EMA) were positive (Strouhal & Němečková, 2004).

IMPACT OF THE TUMOUR ON IMAKHETKHERRESNET'S PATHOGRAPHY

The neurilemmoma must have been growing in Imakhetkherresnet's sacrum for several years. Initially she did not know of its existence, that is, until it became painful. When it increased in size, it inevitably compressed a part of her sacral plexus, with its lobes obstructing three sacral foramina anterior and posterior (right and left S_2 as well as left S_3). At that point she must have experienced paraesthesiae and pain in the lower half of her body. She also could have experienced motoric problems in the pelvico-trochanteric, gluteal and leg muscles. However, the remaining unaffected constituents of the sacral plexus could have ensured her mobility, as shown by her well preserved skeleton without signs of osteoporosis. It is likely that she could walk and was not confined to bed. Most probably she did not die of this tumour.

CONCLUSIONS

Imakhetkherresnet's neurilemmoma, originating in Schwann cells of the myelin nerve sheath, was first established macroscopically and was confirmed by standard radiography and CT imaging of the sacrum. The latter truthfully reflects the shape of the tumour. The diagnosis was definitely confirmed by histological examination and by two positive immunohistochemical tests.

In palaeopathology, a diagnosis with as high a degree of certainty as ours is rare and so is the sacral location of neurilemmoma. In view of the fact that no more than 50-70 modern cases have been published by now (Fechner & Mills, 1993; Strouhal et al. 2003, Strouhal & Němečková, 2004), the finding of our case seems exceptional. In fact, this is the first case of neurilemmoma described in palaeopathological or medical historical literature.

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SAŽETAK

Imakhetkherresnet je pokopana u dobi od 35 do 45 godina u južnome hodniku neoskvrnute grobnice svećenika Iufaae at Abusira (kasna 26. dinastija, 625. g. pr. n. e.). Grobnicu su iskapali istraživači češkog Instituta za egiptologiju od 1994. do 2004. Morfometrijska, genetska i epigenetska svojstva povezuju je rodbinski s Iufaaom, a epigrafska analiza upućuje na to da je bila njegova sestra.

Njezina patografija obuhvaća uobičajene bolesti zuba te osteofitozu kralježnice i degenerativni osteoartritis u ranoj fazi nastanka. Preboljela je spiralni lom kostiju desne potkoljenice.

Na sakrumu se nalazi velika šupljina glatkih stijenki, oblikovana pritiskom relativno tvrdoga tkiva. Njezina veličina i režnjast oblik najprije su uočeni makroskopski, a potom i uobičajenim radiografskim postupkom. CT sekcije pokazale su velike šupljine koje su se protezale od kralježničkog kanala do oba foramena na drugome sakralnome kralješku, te do lijevog foramena na trećem kralješku. Makroskopski i radiografski je dijagnosticiran, a histološki potvrđen dobroćudni neurilemom. Po obliku i lokalizaciji, ovo je prvi takav tumor opisan u paleopatologiji i povijesti medicine.

Ključne riječi: patografija, bolesti kostiju i zuba, neurilemom, Abusir, 26. dinastija, Egipat