Case Report: Tophaceous pseudogout in the temporomandibular joint extending to the base of the skull: Crystallography identification by X-ray diffraction and Fourier transform infrared spectroscopy

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Abstract. We report a case of tophaceous pseudogout (i.e., calcium pyrophosphate dihydrate [CPPD] crystal deposition disease) in the temporomandibular joint (TMJ) that extended to the base of the skull. A 38-year-old man was referred to our hospital with mild pain in the right chin and tip of the tongue. Panoramic radiography showed a large calcified mass around the right TMJ. Computer tomography (CT) imaging revealed a large, granular, calcified mass surrounding the right condylar head and extending to the base of the skull. The mass was clinically and radiographically suspected to be a pseudogout lesion. A biopsy specimen was collected under general anesthesia to confirm the diagnosis. The mass histologically contained the deposition of numerous rod-shaped and rhomboid crystals, which suggested tophaceous pseudogout. The deposition was identified as CPPD crystal deposition, based on analysis by X-ray diffraction (XRD) and Fourier transform infrared spectroscopy (FT-IR). These two crystallography methods were useful in confirming the diagnosis of CPPD crystal deposition disease in the TMJ.

Introduction

McCarty et al.[1] first identified calcium pyrophosphate dihydrate (CPPD) crystals rather than sodium urate crystals in the synovial fluid of patients who had gout-like symptoms; they termed the condition “pseudogout.” The term “tophaceous pseudogout” has recently been used to describe lesions that have massive or tumoral CPPD crystal deposition. This variant is one of the rarest forms of CPPD deposit disease; however, it is important because it shares a histological and clinical resemblance to cartilaginous t tumors.[2, 3] We present a case of tophaceous pseudogout in the temporomandibular joint extending to the base of the skull. We also present images of tophaceous pseudogout in X-ray diffraction (XRD) and Fourier transform infrared spectroscopy (FT-IR).

Case report

A 38-year-old man was admitted to our hospital for diagnosis and treatment of mild pain in the right chin and tip of the tongue. He first noticed mild pain in the right chin 2 months before admission. He had received root canal treatment in his right lower second molar at a neighboring dentist. However, the pain in the area did not subside and he was admitted to our hospital.

His medical history included: hyperlipidemia, gout, diabetes, and hypertension, however, these conditions were well controlled with medication. It is interesting that he had been playing sumo wrestling since he was16 years old. He was a high school teacher and coach of the sumo club. As part of his role as a sumo coach, he always received the students' tackles (i.e., *ukemi* ["passiveness"]) on the right half of the body including his right chin.

Clinical examination showed an obvious preauricular swelling on the right side, which was tender to the touch. His intrinsic mouth opening was limited to 38 mm.

However, he did not have any symptoms that affected his day-to-day function and quality of life.

Computed tomography (CT) scans revealed a calcified mass around the right TMJ, but it was not continuous with the mandibular condyle (Fig. 1A). The calcified mass pressed on the temporal bone and an erosive bone resorption had occurred at the base of skull (Fig. 1B). The CT views of the right TMJ showed limited opening positions due to the right condylar opaque mass. In addition, the calcified mass around the mandibular condyle gained mobility and changed shape on translating the joint from the open to closed position (Fig. 1C). These CT views led us to suspect a pseudogout crystal mass. However, we were unable to completely confirm that the lesion was not a tumor. Because the lesion extended to the base of skull, we discussed the case with neurosurgical specialists. As a result of the discussion, we performed a biopsy of the mass under general anesthesia to confirm the diagnosis. An intraoral incision was created on the mucosal membrane at the anterior margin of the mandibular ramus. The periosteum of the lateral surface of the mandibular ramus toward the processus coronoideus was detached. After reaching the mandibular notch, a significant amount of

intracapsular calcareous material, which appeared chalky or “grist-like” was between the processus coronoideus and the articular processus (Fig. 2A). The biopsy samples were gently removed from the inside of the mass by using a sharp surgical spoon (Fig. 2B).

One-half of the biopsy specimen was immersed in 10% formalin solution and used for the pathological examination. The examination of the specimen revealed that the mass contained numerous deposits of rod-shaped or rhomboid crystals. The background of the crystals was cellular fibrous tissue and foreign body-type giant cells (Fig. 2C). These views suggested CPPD crystal deposition

For the differential diagnosis, the remaining half of the specimen was examined by XRD and FT-IR spectroscopy using the potassium bromide (KBr) disk method. The XRD profile was recorded using a D8 Advance diffractometer (Bruker AXS, Karlsruhe, Germany) diffractometer operated under 40 kV-40 mA acceleration using copper-potassium alpha (Cu Kα) radiation. The patterns obtained by XRD showed that nearly all peaks corresponded to those of CPPD (JCPDS-ICDD-PDF#00-041-0488), although some peaks with an unknown pattern were also observed (Fig. 3A). The FT-IR

spectrum was recorded on an FT/IR-6700 spectrometer (JASCO, Tokyo, Japan) spectrometer at a 4 cm^{-1} resolution. The FT-IR spectrum of calcium pyrophosphate tetrahydrate (CPPT) crystal has been reported,[4] and it is similar to the FT-IR spectrum obtained in this study (Fig. 3B). In brief, the following were recorded: the O-P-O bending vibrations at 509 $cm^{-1}$and 568 cm^{-1}; P-O stretching vibrations at 923 cm^{-1}, 990 cm^{-1}, 1037 cm^{-1}, and 1089 cm^{-1}; O-H plane-bending vibration at 1659 cm^{-1}; and broad peak around 3300 cm^{-1} due to the absorption of water. These data provided further support for the histological diagnosis of CPPD deposition disease, including tophaceous pseudogout.

After the diagnosis was confirmed, we discussed the treatment plan with the neurosurgeons. The patient did not have any disruption to his daily function or everyday life as a result of the lesion aside from the mild pain in the joint, and the lesion was not neoplastic. Based on the situation and the clinical and pathological findings, the neurosurgeons recommended not to resect it because severe neurosurgical dysfunctions could occur. The patient ultimately did not desire to have the mass resected. He continued to coach sumo wrestling, but decided that he should not receive student

tackles to the right chin in the future. Three years after the first visit, the mass showed virtually no change in size.

Discussion

CPPD crystal deposition occurs in a generalized or local pattern.[5] Generalized CPPD crystal deposition is often associated with medical conditions, such as hyperparathyroidism, chronic gout, renal failure, hypomagnesemia, and hypophosphatemia.[6] In this case report, the patient had gout, but the condition was controlled with medication. This finding suggests that CPPD crystal deposition may occur, even if gout is well controlled. Local CPPD crystal deposition occurs secondary to trauma and results in tissue degeneration or necrosis.[5] Our patient had been participating in sumo wrestling for a long time and always received tackles to the right cheek. It is possible that there is an association between the patient's history of participating in this sport and the force exerted on the right cheek and his development of tophaceous pseudogout in the TMJ.

The term "tophaceous pseudogout" has also been used to describe lesions that have massive or tumoral CPPD crystal deposition. Tophaceous pseudogout more frequently occurs in large joints such as the knees, hips, wrists, and pubic symphysis.[3] However, it can also affect the small joints, including the TMJ.[7] Tophaceous pseudogout in the TMJ

share clinical and radiographic features with neoplastic disorders, most frequently pain, swelling, and limited opening of the mouth. However, a wide range of clinical symptoms has also been reported such as facial pain, otalgia, trismus, delayed healing, preauricular tenderness and swelling, and joint destruction.[5, 8~17] In these cases, clinicians usually consider neoplasms as a differential diagnosis. Potential diagnostic possibilities include fibrous dysplasia, synovial chondromatosis osteochondroma, chondroblastoma, and chondrosarcoma.

In this patient, CT scans revealed a large calcified mass around the right TMJ, but the mass had no continuity with the mandibular condyle or other neighboring bones. In addition, it gained mobility and changed shape on translating the joint from open to closed position. These views suggested that the calcified mass was not fibrous dysplasia. Synovial chondromatosis was also ruled out because it is characterized by the formation of small, multiple, metaplastic nodules of cartilage there are generally within the joint space. It is difficult to differentiate tophaceous pseudogout from malignant tumors such as osteochondroma, chondroblastoma, and chondrosarcoma. The calcifications are usually in these tumors, although the calcifications are generally heterogeneous in CT

scans. Furthermore, the contour of the tumor is relatively unclear. These characteristics are different from the characteristics in this case; therefore, we suspected tophaceous pseudogout. However, the radiographic features of tophaceous pseudogout of the TMJ are nonspecific, and we were unable to completely rule out other diseases. Therefore, we performed a biopsy to make a definitive diagnosis. There is one report that could not make a definitive diagnosis by fine-needle aspiration;[5] therefore, we planned an open biopsy.

Pathological diagnosis of the specimen revealed that the mass contained numerous deposits of rod-shaped or rhomboid crystals, suggesting tophaceous pseudogout. In 1981, Martel et al.[18] stated that evidence of CPPD crystals using a polarizing microscope and chemical analysis was necessary for a diagnosis of CPPD deposition disease. Many reports of CPPD deposition disease have been diagnosed using electron probe microanalysis.[8,9,13,14] Electron probe microanalysis can detect the ratio of calcium and phosphorus components in a specimen; however, this method cannot identify the crystal structure. On the other hand, the use of methods such as XRD and FT-IR allows the identification of the crystal structure and element. Therefore, in this

patient we chose to use XRD and FT-IR, which led to a definitive diagnosis. X-ray diffraction is most widely used to identify unknown crystalline materials such as minerals and inorganic compounds. Electron clouds surrounding an atom in a crystal structure tends to diffract X-rays. Therefore, XRD measurement gives XRD patterns consisting of the intensity and diffraction angle. The obtained XRD pattern can be identified by checking the powder diffraction files (PDF) of the International Center for Diffraction Data (ICDD; Newtown Square, PA). For most patients, the identification of the obtained XRD pattern is performed by using the function of “peak search” in the application installed in the PC attached to the XRD machine. Fourier transform infrared spectroscopy is used to obtain the infrared absorption spectrum of the specimen. Some infrared radiation is absorbed by the specimen, whereas some radiation passes through. As a result, the spectrum represents the molecular absorption. When hydroxyapatite [$Ca_{10}(PO_4)_6(OH)_2$] is measured by FT-IR, molecular absorption such as P-O bending and stretching vibration are detected in the resulting spectrum.

Treatment of CPPD crystal deposition in the TMJ varies, according to the extent of clinical findings and the symptoms of the patient. Painful but nonexpansile cases of

CPPD deposition may be treated by a nonsurgical approach using nonsteroidal anti-inflammatory medication or by conservative arthrotomy.[9] On the other hand, surgical excision is the main treatment for tophaceous pseudogout lesions.[5,19]

After the excision of the mass, symptoms were relieved and the patient's ability to open his mouth increased. Surgical excision remains the best therapeutic option for the management of tophaceous pseudogout. However, in this patient, CPPD crystal deposition was substantial, and pressed onto the temporal bone and extended to the base of skull. Surgical excision of the mass may have caused severe neurosurgical dysfunctions. The patient decided not to be operated on. Furthermore, he continued to coach sumo, but decided not to receive any tackles to the right chin at in future.

In conclusion, we reported a rare case of tophaceous pseudogout characterized by CPPD crystal deposition lesions in the TMJ that was managed via nonsurgical treatment (which is contrary to the treatment protocol in previous reports[5,19]). Three years after the first visit, the size of the mass showed nearly no change. Management should involve careful monitoring of the progress of the lesion.

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Competing interests

None.

Ethical approval

Not required.

Patient consent

The patient gave informed consent for the use of his information in this study.

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Figure legends

Figure 1. Computed tomography (CT) imaging shows a large calcified mass around the right temporomandibular joint (TMJ). (A) The axial CT scan shows a ring-shaped calcified mass around the condylar process of the right TMJ. The mass is not continuous with the mandibular condyle. (B) The coronal CT reveals a calcified mass in the joint space. Bone resorption and thinning of the middle cranial base are present and the lesion appears to extend into the middle cranial fossa. (C) The sagittal CT scan image of the right TMJ. The calcified mass limits the condylar head movement.

Figure 2. We performed an intraoral biopsy. (A) The intraoperative view of the mass in the temporomandibular joint (TMJ) region shows the deposition of a “chalky” calcified material (arrow). The arrowhead points directly to the right coronoid process of the mandible. (B) The specimen appears white and “chalk-like.” (C) Histological examination of the specimen shows deposits of crystals (indicated by the arrowheads) in fibrous tissue (stained by hematoxylin-eosin). The crystal deposition consists of

rod-shaped and rhomboid crystals, which are surrounded by foreign body-type giant cells (denoted by “G”) and fibroblasts (denoted by “F”).

Figure 3. The X-ray diffraction (XRD) pattern and Fourier transform infrared spectroscopy (FT-IR) spectra of the specimen. (A) The XRD pattern shows that most peaks correspond to those of calcium pyrophosphate dihydrate (CPPD) (JCPDS-ICDD-PDF#00-041-0488). The “▽ ” symbols indicate the intrinsic peaks of CPPD. (B) The FT-IR spectrum is similar to that of CPPD.

Figure 1.

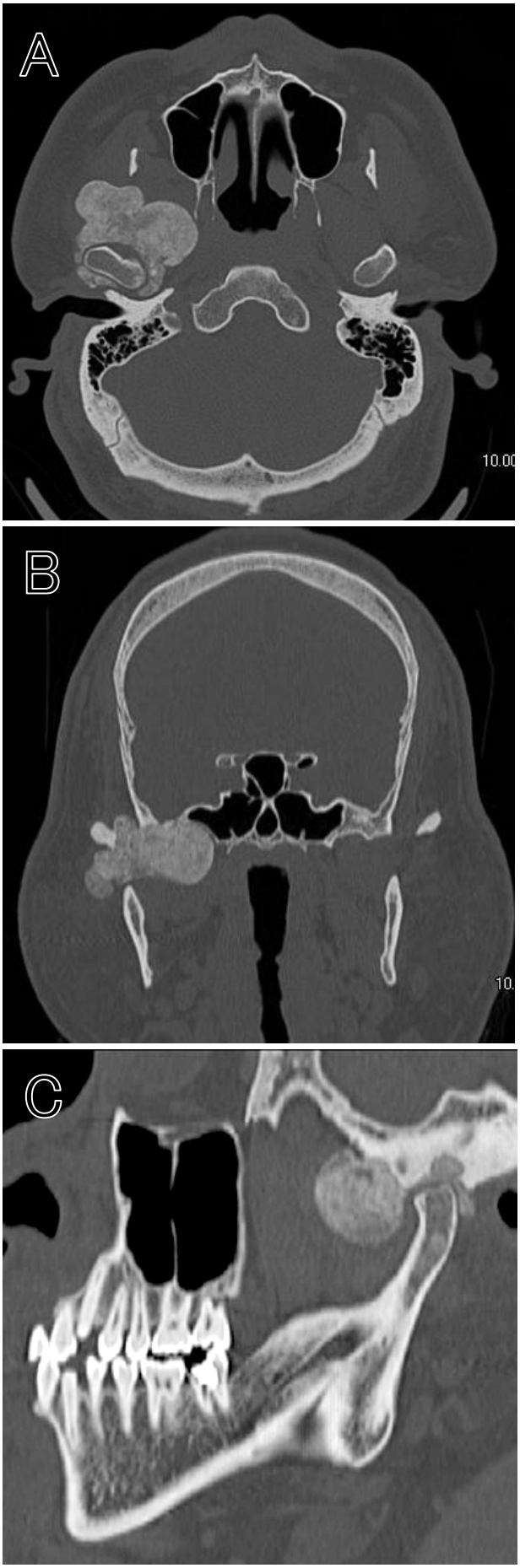


Figure 2.

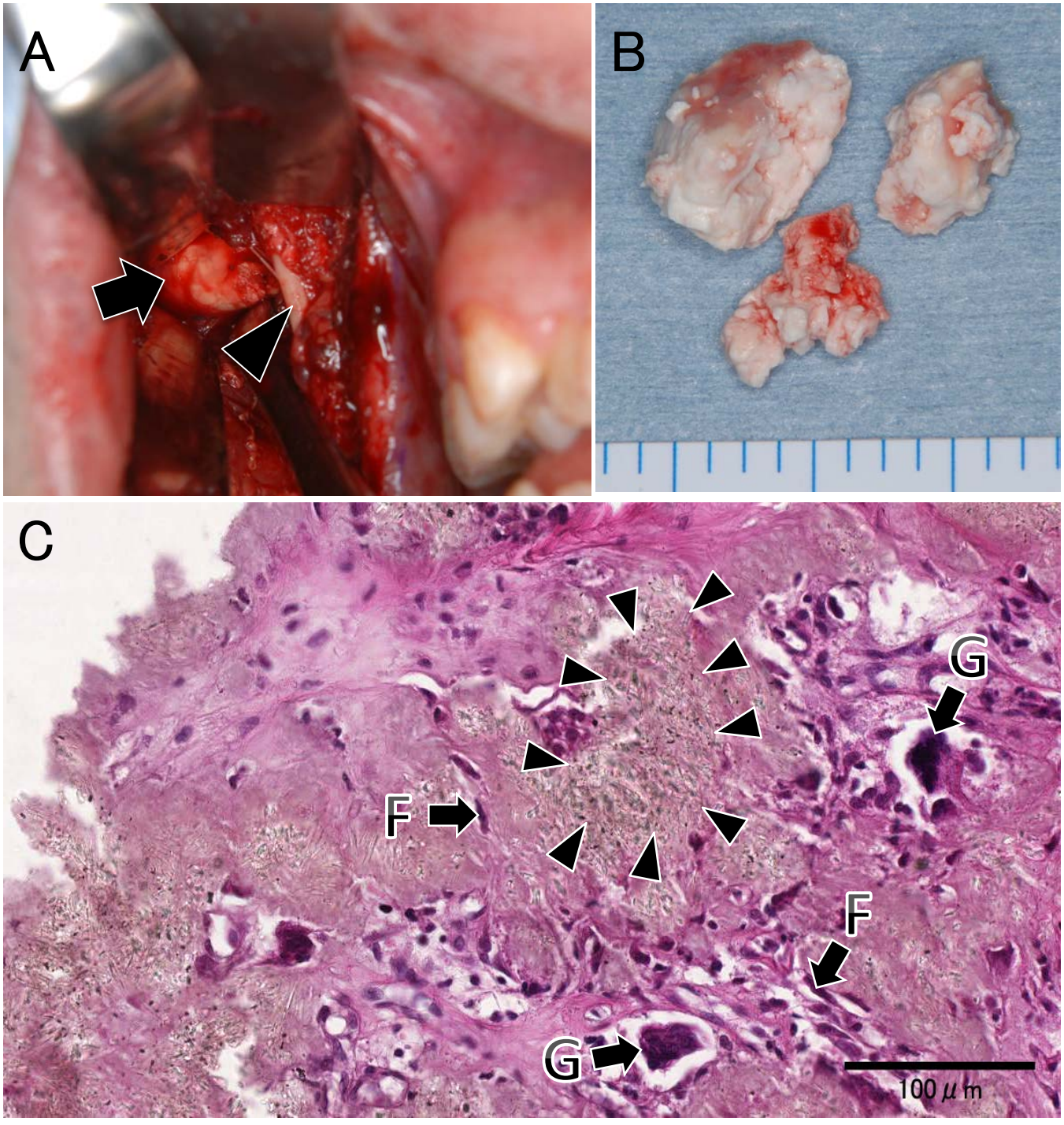
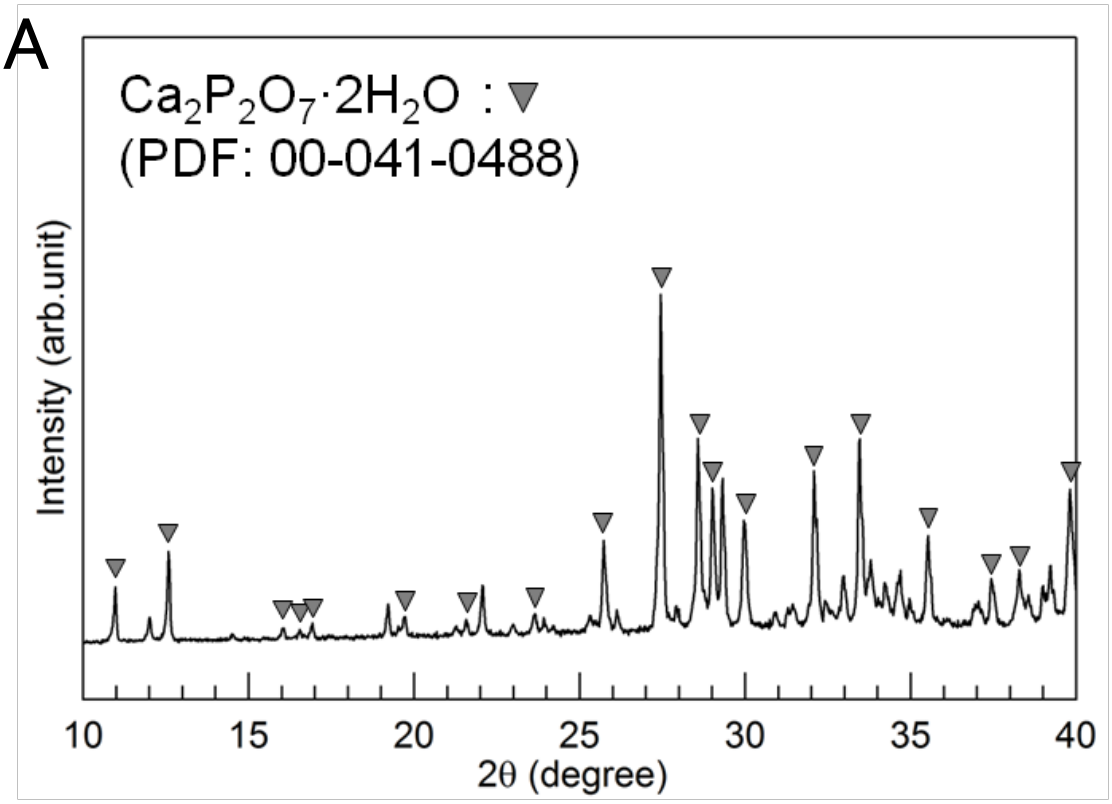


Figure 3.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Intensity (arb.unit)
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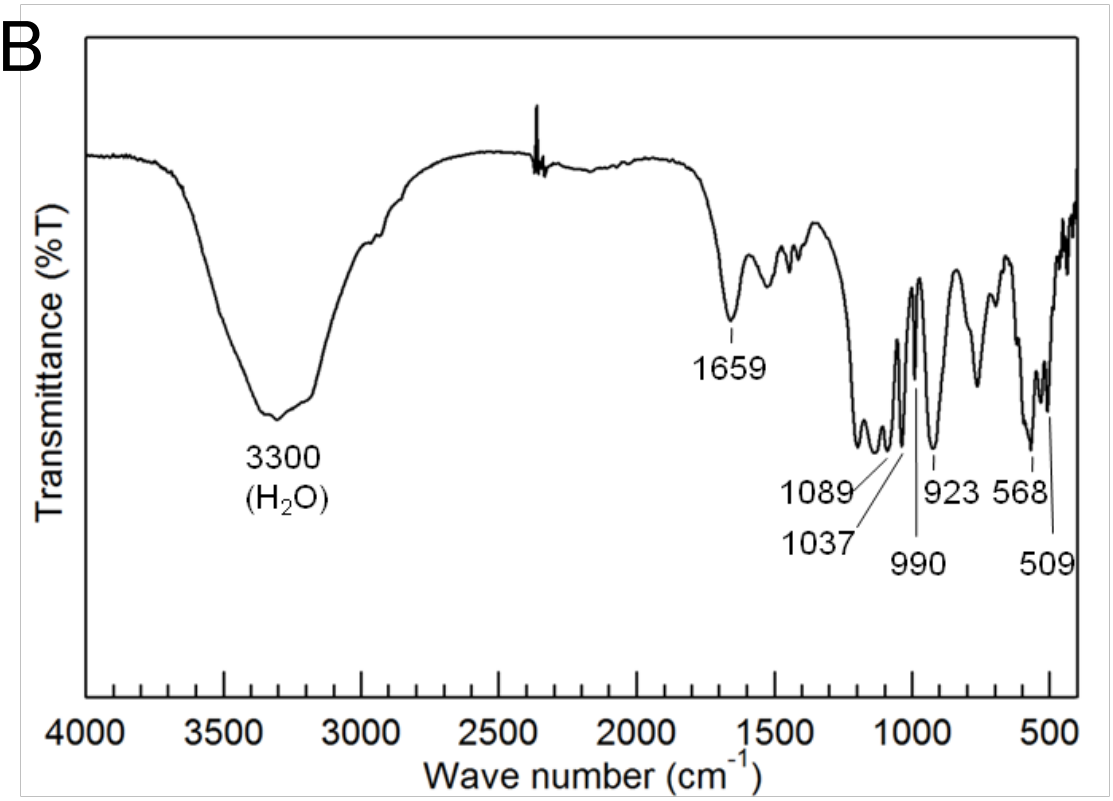
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Wave number (cm^{-1})