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Surgical Neuropelveology: Laparoscopic Nerve-Sparing Excision of A Pelvic Schwannoma Guided by Electrophysiological Study

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1. Abstract

1.1. Background

Schwannomas are benign tumors derived from the glial cells of peripheral nerve sheaths and are usually composed of well-differentiated Schwann cells. Retroperitoneal location of schwannomas are very rare, accounting for less than 2%1. Although mainly benign with a low operative mortality rate, schwannomas carry significant risk with high morbidity and surgical challenges because of their late nonspecific presentation as well as their size and anatomical location.

1.2. Observations

The authors describe the management of a woman with a 4-centimeter schwannoma arising from the right nerve root of S2 of her pelvis, presenting as a pauci symptomatic lesion. Treatment involved total laparoscopic excision with the assistance of probes for neurophysiological study of nerve pathways, aiming to minimize potential nerve injuries.

1.3. Lessons

We confirmed the feasibility of laparoscopy in the management of retroperitoneal gynecological peripheral nerve neoplasms, despite the lack of literature, due to the rarity of this tumor. We also aimed to demonstrate how the use of neurophysiological techniques, which are rarely employed in gynecological settings, can enable the performance of delicate surgical procedures with a higher degree of safety and precision, minimizing the possibility of iatrogenic damage.

2. Introduction

Schwannomas are benign tumors derived from the glial cells of peripheral nerve sheaths and are usually composed of well-differentiated Schwann cells. A very rare location for schwannoma is in the retroperitoneal areas (less than 2%) [1]. Here, we report a case of a patient with a 45x43mm schwannoma arising from the nerve root of S2 on the right side, presenting as a pauci symptomatic lesion found on ultrasound. Treatment involved total laparoscopic excision with the assistance of probes for neurophysiological study of nerve pathways, aiming to minimize potential nerve injuries. Although mainly benign with a low operative mortality rate, schwannomas carry significant risk with high morbidity and surgical challenges because of their late nonspecific presentation as well as their size and anatomical location.In the existing medical literature few laparoscopic resections of retroperitoneal schwannomas have been documented, and we believe ours to be the first case performed where the surgical gesture was guided by the electrophysiological study of nerve conduction in order to avoid iatrogenic lesions.This article extensively discusses technical details of the procedure.

3. Case Report

A 53 year old, Caucasian, obese (BMI 31), pre-menopausal women presented at our hospital with a recent finding on gynecological ultrasound performed at another facility of a 4^{cm} solid retro uterine lesion of uncertain nature. Patient was symptomatic for chronic pelvic pain radiated to the right thigh. A magnetic resonance was

performed and a rounded lesion of 40mm in the presacral region with heterogeneous signal intensity, showing close continuity with the sacrum and the left sacral foramens, highly suggestive of schwannoma, was described. (Figure1a and Figure 1b).The ultrasound scan in our institution showed a hypoecho roundish mass of 44x34mm, with regular external borders, located between the rectum and the posterior wall of the uterus with no color flow (Color Score 1) at the Power Doppler study. The mass was in close contiguity with the left posterolateral wall of the uterus while maintaining a cleavage plane. No pain was evoked at the targheted pressure with transvaginal probe(Figure 2a and Figure 2b). The patient was also visited by a neurosurgeon that excluded any sensitive or motor disturbance and any other neurological symptoms. After detailed discussion with the patient about the risks of sacral nerve injury, due to the uncertain nature of the lesion and its symptoms, a laparoscopic excision was scheduled, with the support of the intraoperative neurophysiological monitoring (IONM)2 that lead us to real-time detection of potential nerve damage during surgery.

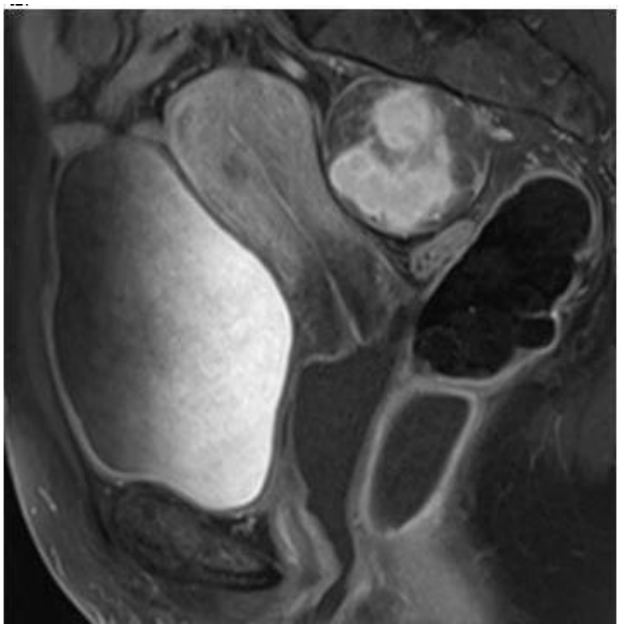

Figure 1a: Sagittal MRI scan showing the tumour.

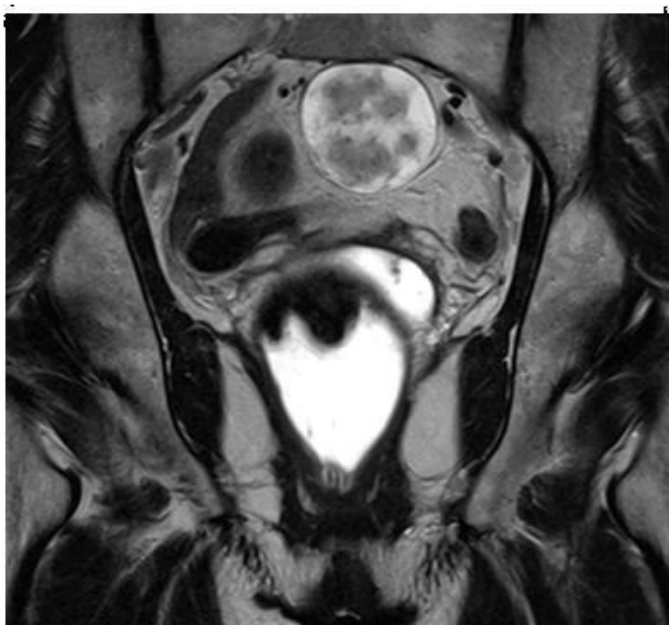

Figure 1b: Coronal view.

Figure 2a: Transvaginal ultrasound showing the lesion**.**

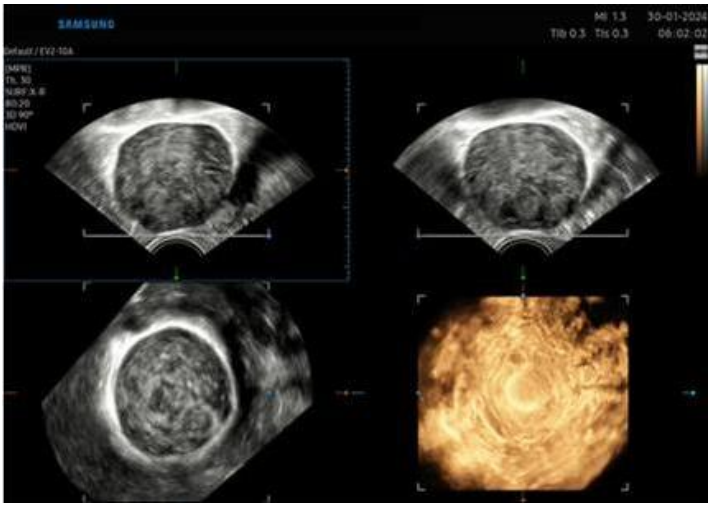

Figure 2b:3D reconstruction of the lesion.

4. Surgical Technique

4.1. Patient Was Positioned in Supine, Gynecological Position, and A Urinary Catheter Was Inserted

Rocuronium bromide Esmeron 50–150 mg was administered intravenously as a short-acting muscle relaxant to facilitate endotracheal intubation. No paralytic agents were subsequently administered and general anesthesia was induced by total intravenous anesthesia since evoked potentials and EMG waves in the designated muscles had to be recorded. Remifentanil Ultiva and propofol Fresofol were used in several combinations to initiate and maintain the general anesthesia. During anesthesia, body temperature, direct radial artery pressure, pulse rate, oxygen saturation, and end-tidal carbon dioxide concentration were continuously monitored. The patient was kept normothermic and normotensive.The surgical procedure started with supraumbilical open laparoscopy, introduction of a 10-mm trocar, and insufflation to an intra-abdominal pressure of 20 mm Hg. After exploration of the abdominal cavity, 5 mm ancillary trocars were positioned as illustrated in Figure 3. A 3D Karl Storz 4k camera was used. The patient was placed in the steep head-down position and the small bowel swept into the upper abdomen to aid visualization of the operative field. To allow exposure of the pelvis, a uterus manipulator was placed.

Figure 3: Trocar Placement.

4.2. The Tumor Was Visible on The Left Side of The Pelvis Trans Peritoneally

The dissection was begun at the sacral promontory, mobilizing the rectum away from the left side of the pelvis. The peritoneum was then incised on the left pelvic brim to allow the identification of the right ureter, the gonadal vessels and the right ipogastric nerve.

Dissection continued posteriorly preserving the integrity of the capsule and ensuring meticulous haemostasis at all times by means of the harmonic scalpel (Ethicon Endosurgery, Cincinnati, USA). The sacral nerve roots were identified confirming that the tumor was coming from S2 (Figure 4).Electromyogram and evoked potentiometer were used for monitoring, and concentric electric needles were inserted into the innervated muscles to record the procedures. The surface electrode was used to stimulate different levels of the tumor-involved nerve and the conduction velocity and potential amplitude recorded (Figure 5). Mapping was conducted with a monopolar ball tip probe, and stimulation was initiated at intervals of 0.1 ms from a voltage of 4 mA, which was increased to 25 mA. During tumor resection, nerve stimulation was frequently used to stimulate the nerve or suspicious capsule to distinguish whether they were safe or not.The capsule of the mass, corresponding to the epineurium, was incised to expose the schwannoma at the point indicated by the electrophysiological study, at the site where the nerve conduction showed the least impulse (Figure 6). In the process of tumor resection, one of the most important steps was to make sure the relationship between the nerve and schwannomas.Cautious enucleation in contact with the schwannoma was performed by separating healthy nerve fascicles (Figure 7). Enucleation was completed by elective coagulation with bipolar forceps and section of the vascular pedicle.

Figure 4: The tumor emerging from the cranial portion of S2.

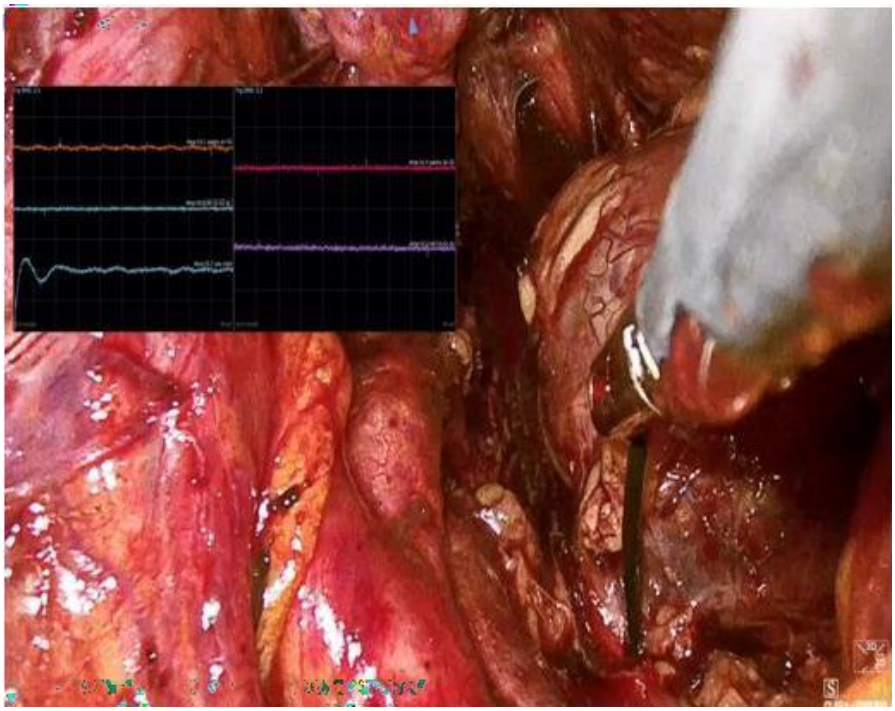

Figure 5: Stimulation of the cranial portion of S2 nerve from which the tumor derives.

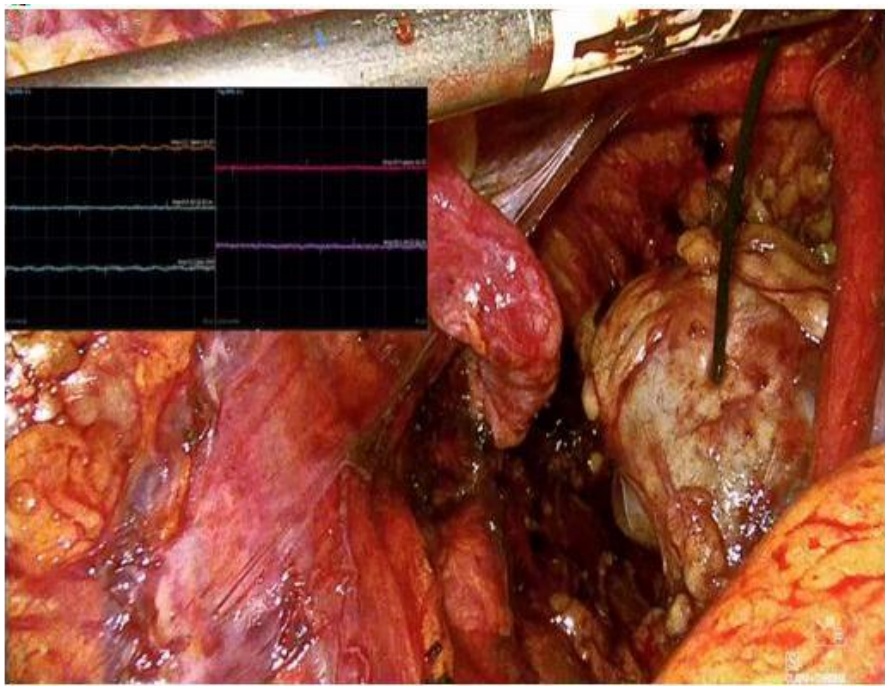

Figure 6: Stimulation of the capsule of the tumor where no signal was recorded, showing a safe area where to start the incision and dissection.

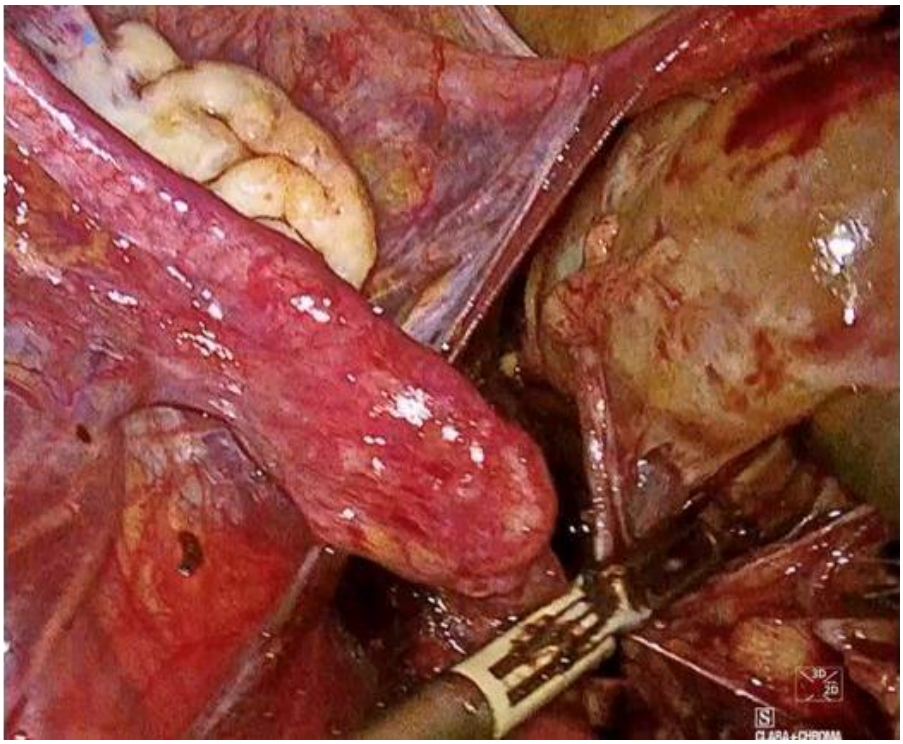

Figure 7: Enucleation of the schwannoma from the healthy nervous bundles.

4.3. The Other Nerve Fascicles Were Preserved

The dissection was completed inferiorly freeing the tumour in its entirety. Tumor was extracted trough a laparoscopic bag via minimal enlargement of the sovrapubic incision, without any spillage (Figure 8). No drains were used.Operative time was 75 minutes and the operative blood loss was lass than 50 ml.Foley was removed on day one. The patency of intestinal tract was obtained in 2p.o. day. Patient was discharged home on postoperative day 2 and had a completely uneventful postoperative period. No change in the sensitivity of the lower limbs, paresthesia or difficulty in emptying the bladder was recorded.At the postoperative control, two weeks after the procedure, the patient showed a complete resolution of her symptoms, no longer suffering chronic pelvic pain.Histologically the lesion was a spindle cell neoplasm without mitotic activity with a strong S 100 protein immunostaining positivity. Smooth muscle actin, desmin, calponin, caldesmon, CD 117, DOG-1, HMB-45 and AE1/AE3 cytokeratins immunostaining was negative. A diagnosis of benign nerve sheath tumor (schwannoma) was so achieved.

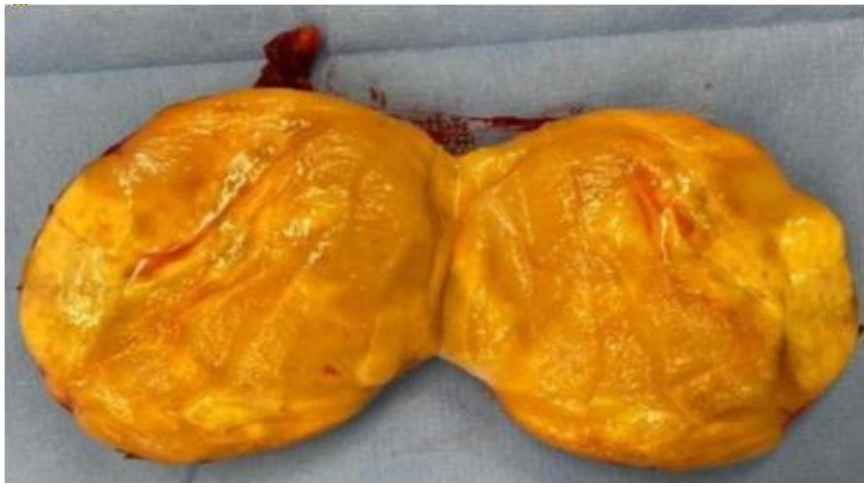

Figure 8: Macroscopic view of the tumor.

5. Discussion

Schwannoma is a benign tumor of the peripheral nerve sheath that arises from Schwann cells and typically manifests as a soft subcutaneous nodule or mass in the trunk. However, diagnosis of schwannoma is frequently delayed for several years as most cases remain asymptomatic initially and grow slowly over time [3].Retroperitoneal schwannoma are very rare45 and those that grow in the retroperitoneum are often malignant (0.3 to 3.2%). Most common locations are cranial nerves (especially the 8th pair) and, in peripheral nerve system, the neck, the mediastinum and extremities. When its onset is combined with Recklinghausen disease (type 1 neurofibromatosis), retroperitoneal development is more common.Symptoms can include sciatica, lower back and abdominal pain, heaviness when micturating or defecating, as well as pelvic heaviness from bladder and rectal compression. Significant motor weakness is less common and is usually noticed with malignant schwannoma 5-6]. As a result of their slow growth rate and anatomic location, pelvic schwannomas usually remain asymptomatic for long time and are either discovered incidentally or as soon as they are sufficiently large to cause a mass effect [7]. This types of tumors are quite difficult to diagnose preoperatively because neither the clinical symptoms nor the radiological characteristics are typical. Ultrasound (US), CT scans and MRI can show a rounded, well defined, solid mass lesion but cannot provide histological certainty. Fine needle percutaneous cytology has been proposed to achieve preoperative diagnosis. However, FNA of soft tissue lesions in the retroperitoneum can result technically difficult and often be unsuccessful especially for large and mixed lesions. [8]

6. Observations

Surgical removal of schwannoma is widely accepted as a routine option [9, 10]. It is generally believed that this kind of tumor can be easily enucleated from the nerve stem without further neurological deficits [11].However, due to long-term compression of the tumor, these nerve bundles usually became small, thin, transparent or translucent, and difficult to recognize. It has been a great challenge for us to recognize and protect these small nerve bundles against nerve injury and for this reason we resorted to the use of electrophysiological study of nerve transmission with the aim of minimizing iatrogenic nerve injuries.IONM is generally used in surgeries that carry a risk of injury to the central nervous system during the procedure, such as brain tumor removal, cerebrovascular surgery, or spinal cord surgery [12]. The main modalities of IONM are evoked potentials, spontaneous EMG, and triggered EMG [13]. These modalities should be comprehensively interpreted to detect neural injury and determine the state of the patient. Spontaneous EMG represents the electric signals from the muscles recorded passively, whereas triggered EMG represents the signals induced by the direct stimulation of nerve or hardware; therefore, they can provide immediate and reliable information about any nerve injury to the surgeon [14]. The choice of surgery can be an anterior, posterior, or a 360 approach in which spinal instrumentation is considered in severe bony destruction. The choice of approach should depend on tumor size and which anatomical compartments it transgresses, i.e., whether it is confined to the presacral region or extends more posteriorly into the spinal canal.

Due to the location of the tumor and the absence of bone erosion, we opted for an anterior approach to obtain a complete and safe resection. Schwannomas are usually solitary, well circumscribed and encapsulated tumors. Due to these characteristics they are easily dissected from adjacent tissue, which makes laparoscopic resection possible. Laparoscopy might also facilitate dissection due to magnification of the anatomic elements in the narrow pelvis.

The literature indicates that laparoscopy excision of these neural tumors should be the therapy of choice [15,16]. Collected retrospectively in 2023 clinical data of 38 patients with lateral pelvic schwannomas [18], treated laparoscopically and 20 treated with open surgery, demonstrating the advantages of the minimal invasive approach: most notably small incisions, less blood loss, as well as potential benefits of neurological function [17].

The intraoperative neurophysiological monitoring (IONM) has allowed us to guarantee the functional integrity of nerves, even in a surgery involving the possibility of a peripheral nerve injury [18]. The principal lessons to be drawn from this case were the need for good exposure with the steep head-down position from the start, careful retraction of the rectum to the left to permit a plane to be developed to the left of the retrorectal mass and then a similar approach to the right side of the retrorectal mass. In addition, awareness of the distorted course of the right ureter made possible early recognition and successful preservation of this structure, and once it was out of the operative field, the more adherent plane behind the mass could be developed. This permitted access to the correct plane around the mass and hence, good haemostasis was maintained. Accurate preoperative imaging of the lesion with identification of the S2 nerve root as the point of origin helped to preserve this structure.Despite that, our case confirmed the feasibility of laparoscopy in the management of retroperitoneal gynecological peripheral nerve neoplasms, despite the lack of literature, due to the rarity of this tumor. The laparotomic approach is certainly more traumatic, more painful, requires a longer recovery time, and will increasingly be considered as a salvage option in those few cases where minimally invasive approach is not feasible.The precise, high-definition, 3D visualization of the surgical field in this deep and narrow anatomical area, particularly in obese patients like ours, offered a detailed view, simplifying the task of isolating and navigating the anatomical structures around the tumor. Additionally, the use of pneumoperitoneum had facilitate the creation of an optimal dissection plane, thereby reducing the likelihood of tumor rupture and bleeding.Secondly, we demonstrated how the use of neurophysiological techniques,

which are rarely employed in gynecological settings, can enable the performance of delicate surgical procedures with a higher degree of safety and precision, minimizing the possibility of iatrogenic damage. It is important to stress out that the nerve fibers are part of the tumor and a complete resection is usually impossibile without some microscopic neurological sacrifice. In our patient the blunt dissection supported by the neurophysiological monitoring made the possible neurological damage to be clinically unrelevant.

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