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Syringomyelia: etiologies and aspects in MRI

Rasolohery H[1], Ramarokoto M[2], Rajaonarison Ny Ony LNH[3], Andrianah GEP[4], Ahmad A[5]
[1-5] Imaging Medical Center, Hospital Joseph Ravoahangy Andrianavalona, Antananarivo, Madagascar

Abstract
Introduction: Syringomyelia are fluid formations in intramedullary spread over at least two myelometers and composed of cerebrospinal fluid. They are rare and caused by various pathologies. MRI is an essential examination that contributes to the diagnosis. The objectives of our study are to determine the aspects in MRI and the etiology of this pathology in Madagascar.
Methods: A retrospective descriptive study realized between March 2017 and March 2019 about 32 patients with MRI-diagnosed syringomyelia. This study is carried out at the medical imaging center of the Hospital Joseph Ravoahangy Andrianavalona Madagascar. The parameters studied were the demographic profile of patients, the clinical symptomatology, the MRI aspects and the etiologies of syringomyelia.
Results: We observed a female predominance with a sex ratio of 0.6 and an average age of 40.81 years. The syringomyelic cavities averaged 3.2 mm (2 to 10 mm) and located mainly in the cervico-dorsal level (40.6%). Tumors were the main etiologies with 37.5% (12 patients), followed by Chiari disease 28.12% (9 patients), myelitis 15.62% (5 patients) and traumatism 6.25% (2 patients). Idiopathic forms accounted for 12.5% (4 patients). The tumoral syringomyelia are developed upstream and limited to the same floor as the tumor. The syringomyelia associated with Chiari's disease have variable size and morphology but localized notably at the cervical and dorsal level. The syringomyelic cavities of inflammatory origin are multicavitary and extensive.
Conclusion: Intramedullary tumors are the main causes of the formation of syringomyelic cavities in Madagascar. The aspects and morphology of the syringomyelia in MRI are polymorphic and vary by etiology.

Keywords: syringomyelia, MRI medullar, tumor, myelitis, Chiari disease

1. Introduction
Syringomyelia are abnormal formations of fluid cavities within the spinal cord, which can progressively increase. It is a rare and chronic disease whose incidence is 8.4 per 100,000 habitants in occidental countries [1]. In Madagascar, according to our knowledge, no study on this pathology has been carried out before. It can manifest by various symptoms [2, 3]. The medullary MRI allows to make its diagnosis. The objectives of this work are to determine the MRI aspects of syringomyelia and its etiologies.

2. Materials and methods
Descriptive and analytical retrospective study of spinal MRI with the medical records of 32 patients with syringomyelia. This study is carried out at the Imaging Medical Center of Hospital Joseph Ravoahangy Andrianavalona over a period of two years (March 2017 to March 2019). The parameters studied were age, gender, clinical symptomatology of patients, as well as aspects of MRI lesions and etiologies of syringomyelia. MRI was concerned all-marrow with T1 and T2 sagittal sections, T1 and T2 axial sections centered on medullary lesions, STIR sagittal sections and 3-planes sections after gadolinium injection.

3. Results
We found a female predominance with 19 women and 13 men, a sex ratio of 0.6. The median age was 50.81 years with extremes of 12 and 71 years.
Clinically, all patients were symptomatic, but the clinical presentation was diverse. Neuropathic pain predominated in 23 patients (71.87%) and evoluted in chronic way. Nine patients (28.87%) presented motor deficits of varying degrees.
On the MRI, the syringomyelic cavity is signal identical to the CSF in hyposignal T1 and hypersignal T2. It was unicavitary in 71.87% (23 out of 32 patients) and multi-cavity in 28.12%. For thickness, the syringomyelic cavities measured from 2 to 10 mm with an average of 3.2 mm in diameter. In 40.6% (13 cases), they are located in the cervico-dorsal, cervical, dorsal and lumbar regions respectively in 12.5% (4 cases), 21.8% (7 cases), 18.7% (6 cases).
Tumors were the most common causes of syringomyelia in this study, observed in 37.5% (12 patients). Ependymoma was the most common. It was constated that the tumoral syringomyelia were small, being limited to the same stages as the tumors involved (Figure 1). In this category of etiology, most patients have been deficient.
Syringomyelia associated with Chiari diseases accounted 28.12% (9 of the 32 cases). They were all unicavitary while the extent and size were variable. In one patient, the syringomyelic cavity laminated completely the marrow (Figure 2).
Inflammatory causes were third in 5 patients or 15.62%. The

diagnosis of myelitis was posed by a bundle of clinical, MRI and therapeutic evidence with parallel regression of the lesion at the MRI control and clinic. Syringomyelia in the course of myelitis were multicavitary and extensive in three patients. They were developing upstream and downstream of the causal lesion that frequently sat in the dorsal region (Figure 3).

Syringomyelia without obvious or idiopathic form accounted for 12.5% (4 cases). Cervical-thoracolumbar spinal MRI did not find any abnormality that could explain the formation of syringomyelic cavities. They are located mainly on the dorso-lumbar floor and were unicavitary.

Traumatic causes were by far the most common in our study. The two cases observed (18.75%) are the result of vertebromedullary traumatism treated surgically. The fluid cavities were developed upstream and downstream of the surgical approach.

4. Discussion

Data from the literature on syringomyelia shows discrepancies in terminology and classification [4]. However, the authors were unanimous on the fact that not all intramedullary liquid formations are syringomyelia [1, 5]. It is differentiated from central channel dilatation, intramedullary tumor cysts, myelomalacia and ependimoglial cysts [5]. The main differential diagnosis to be eliminated is the persistence of the central ependymal canal, which is usually <3mm in diameter that they are considered pre-syringomyelia state [1].

Syringomyelia are fluid cavities that increase over time, extending in diameter at the top and bottom as well as in width [6]. The presence of a cause most often at the level of the cranio-cervical junction is also an important element contributing to the diagnosis.

Clinically, the manifestation is presented mainly by neuropathic pain. The more syringomyelia are important, the more the symptoms are increased. One study showed the correlation between syringomyelia and the presence and intensity of neuropathic pain [7]. In our case, patients with idiopathic syringomyelia had neuropathic pain of chronic evolution. For syringomyelia associated with myelitis, clinical signs and MRI lesions improved or decreased with treatment.

Syringomyelia associated with tumoral and degenerative causes are asymptomatic [8, 9]. Clinical signs are related to tumor compression or disc disease. In our case, patients with tumors are mostly deficient.

The most common causes of syringomyelia reported in the literature are mainly craniocervical junction abnormalities, which cause a change in CSF flow [9, 10, 11]. It is essentially type 1 Chiari disease. It represents 35.4% of cases in the Jork Klemkap study [9]. If most literature studies reported the mechanisms and causes of the formation of syringomyelia; few studies have talked about MRI aspects of syringomyelia. The morphology of syringomyelic cavities associated with Chiari disease is variable. Toshikata Seki and his team [12] classified into four types: deviated, expanded, central and partitioned. They were found that 11 of the 24 patients with Chiari disease associated with syringomyelia, the cavities are deviated. syringomyelia were beginning at the level of the cervical stage and extend downwards. In our case, one was enlarged completely rolling the marrow.

The tumoral causes are in second place representing including intra and extra-medullary tumors [9]. Intramedullary tumors are often associated with syringomyelia in relation to extramedullary tumors [13]. The pathophysiological mechanism is not clear. In the Jork study, 152 patients with intramedullary tumor versus 92 patients with extramedullary [9]. They were often developing upstream of the tumor and confined to the same floor as the tumor [13]. This aspect is like our results.

In our study, tumors were the first causes before Chiari's disease; this discrepancy with the literature could be explained by the fact that in our country, MRI is not a technique that is still easily accessible and that spinal tumors are much more clinically obvious requiring further investigation such as this technique.

For myelitis, the mechanism of development of the syringomyelia cavity is related to an accumulation of vasogenic edema and is formed at the acute stage of myelitis [14]. Syringomyelic cavities are non-communicating -. Similarly, in our case, all syringomyelia of inflammatory origin were multicavitary. The cause and effect relationship are established by the development and resolution of syringomyelic cavities and myelitis [14].

For idiopathic forms, no cause has been identified on MRI. Some authors have reported a pre-syrinx condition that is manifested just by an enlargement of the marrow without cavitation [15]. At this stage, the patient is already symptomatic and after an intervention that attenuates the CSF flow obstruction, the marrow enlargement has disappeared at the control MRI. For this etiology group, the syringomyelic cavity is unicavitary. It can be localized corresponding to an enlargement of the central or extended canal and can cause a neurological dysfunction [16]. In our study, it is mostly localized and sits at the lumbar level. They can be of chance discovery [4]. In our case, all cases were symptomatic with neuropathic pain.

5. Tables and Figures

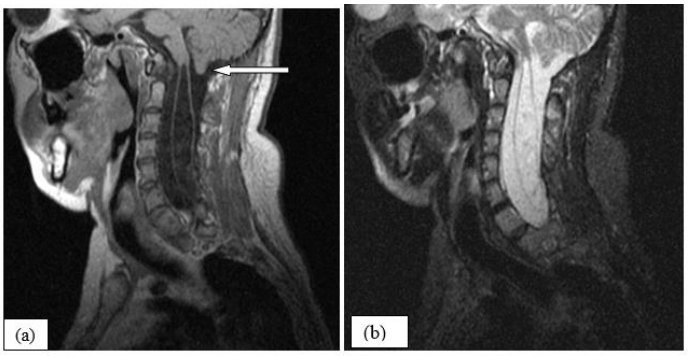

Fig 1: Cervical medullary MRI showing T1 hypointense syringomyelic cavity (a) and T2 (b) hypersignal by protrusion of the cerebellar amygdal: Chiari disease (arrow).

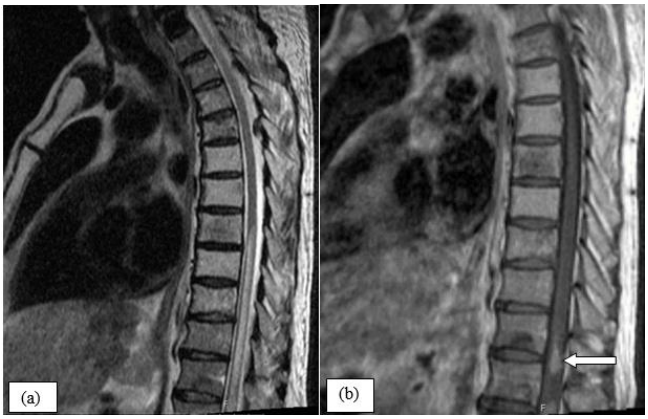

Fig 2: Dorsal medullary MRI showing syringomyelia in T2 (a), tumor lesion with enhancement after gadolinium injection (b): Ependymoma (arrow).

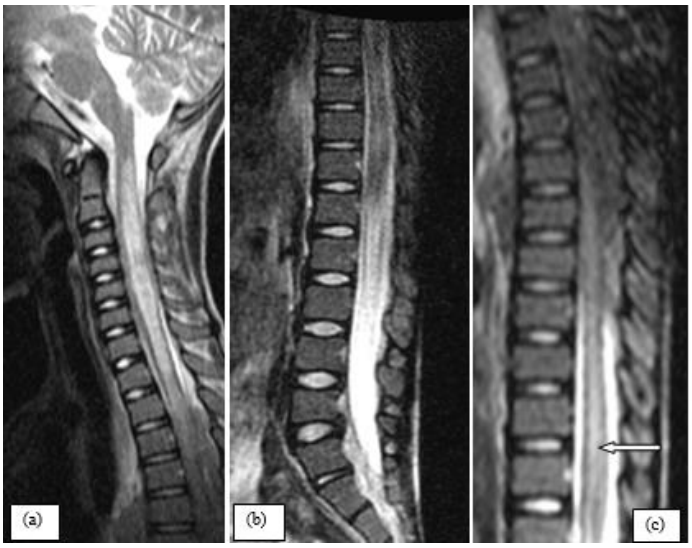

Fig 3: Cervical (a) and lumbar (b) medullary MRI showing syringomyelic cavities in T2 hypersignal; Dorsal medullary MRI (c) showing heterogeneous regular enlargement STIR: myelitis (arrow).

6. Conclusion

Syringomyelia can be idiopathic or secondary to medullary pathologies. In Madagascar, tumors are the main causes of syringomyelia. MRI aspects and morphology are polymorphic and differ in etiology.

7. References

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