Case Report

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Intra thecal baclofen pump as an ad hoc measure for a case of severe tardive dystonia refractory to multiple lesioning

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Introduction and Importance: Tardive dystonia is an infrequent ailment in patient reliant with chronic antipsychotic medication. The front-line envoy in the treatment of this illness is set into motion with oral agents including baclofen, benzodiazepines, and other antispasmodics. Regardless of an extensive therapy, the patients are not able to control of their spasticity/ dystonia. The authors reported a case of severe tardive dystonia treated with baclofen therapy in a patient frigid to medical therapy and multiple lesioning.

Case Report: A 31-year-old female, diagnosed as a case of depressive illness and being managed with neuroleptic medications, who went onto develop tardive dystonia progressively worsening over a 4-year duration. After a comprehensive and meticulous evaluation of her neurological and psychological stratum, globus pallidus interna lesioning was reputed as the best course of action. As intended, staged lesioning was executed bilaterally with a trivial resolution eventually succumbing into recurrence, compelling a repeat lesioning. It was inaptly discouraging to see her crippled with the plight. Determined, not to give upon her, a way out with a baclofen therapy was proposed. A test dose with a 100 mcg of baclofen with an increment up to 150 mcg over a 3-day period demonstrated a promising prospect. On that account, the insertion of the baclofen pump was performed with an outstanding aftermath in her neurological endeavor.

Clinical Discussion: Tardive dystonia is believed to be caused by striatal dopamine receptor super-sensitivity persuaded by the dopamine-antagonizing action of antipsychotic drugs. The first line of treatment being oral agents including oral baclofen, benzodiazepines, and antispasmodics. If the patient suffers from an early-onset primary generalized dystonia, then treatment with deep brain stimulation of the globus pallidus interna is the approved and preferred treatment approach. Recurrence of the symptoms despite of multiple lesioning can be overcome by intrathecal baclofen pump infusion as stated by many research. It is not uncommon to face complications in such a procedure, but the benefits outreach the risk, which makes it a choice of treatment.

Conclusion: The use of a continuous intrathecal baclofen pump for cases with tardive dystonia refractory to conventional therapy, it has been approved as one of the safest and capable procedures.

Keywords: baclofen, implantable pump, intrathecal, movement disorder, tardive dystonia

Introduction

Tardive dyskinesia is a syndrome of purposeless, repetitive, involuntary movements usually involving the mouth, tongue, and face, and sometimes the extremities and trunk[1]. Introduced in 1973, the term tardive dystonia was coined to label a movement disorder that should encompass a spectrum of criteria viz. presence of chronic dystonia, history of antipsychotic drug treatment preceding or concurrent with the onset of dystonia, exclusion of

HIGHLIGHTS

- Tardive dystonia is an infrequent ailment in patient reliant with chronic antipsychotic medication.
- The front-line envoy in the treatment of this illness is set into motion with oral agents including baclofen, benzodiazepines, and other antispasmodics.
- If the patient suffers from an early-onset primary generalized dystonia, then treatment with deep brain stimulation of the globus pallidus interna is the approved and preferred treatment approach.
- Recurrence of the symptoms despite of multiple lesioning can be overcome by intrathecal baclofen pump infusion as stated by many research.
- It is not uncommon to face complications in such a procedure, but the benefits outreach the risk, which makes it a choice of treatment.

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known causes of secondary dystonia by appropriate clinical and laboratory investigations, and no family history of dystonia[2]. Tardive dystonia is an infrequent ailment registered to affect a mere 1.5–2% of patients reliant with chronic antipsychotic medication[3–5].

The front-line envoy in the treatment of this illness is set into motion with oral agents including baclofen, benzodiazepines, and other antispasmodics. Regardless of an extensive array of armaments of therapies, the sufferer grapple to attain a copacetic control of their spasticity/ dystonia[6][7]. Trials with oral baclofen has been reported every so often. Inconsolably, it is acknowledged that only a small fraction of oral baclofen infiltrates into the central nervous system[8]. To contravene to this predicament, the use of intrathecal baclofen was suggested. A remarkable concentration of baclofen was conveyed to the central nervous system in an order of 2–3 times the magnitude greater than those achieved with systemic therapy[7]. Not to mention, this strategy has also been found to be helpful in the treatment of poststroke central pain, dystonia passive to conventional therapy, and persistent vegetative state[9–12]. In recent times, an aggregate of evidence has been disclosed in favor of intrathecal baclofen infusion as an affirmative in the therapeutics of spasticity[7].

We herein, report a case of severe tardive dystonia treated with baclofen therapy in a patient frigid to medical therapy and multiple lesioning. This case report has been reported in line with the Surgical CAse REport (SCARE) Criteria[13].

Case report

A 31-year-old woman with 4-year history of progressive worsening of tardive dystonia who was medicated with antipsychotic prescription as a remedy to her depressive illness. She was so much reliant on the use of neuroleptics, that she started noticing occasional stiffness of her body. At the outset, it was not too bothering and thus she refrained from seeking medical attention. Unfortunately, at the tail end the severity soared up so grievously that she ended up suffering a dislocated Temporo-mandibular joint due to a profound stiffness of her facial musculature. The Burke- Fahn-Marsden cervical dystonia rating on examination was found to be 36[12]. She was then instituted with on oral medications (Benzodiazepines, Oral Baclofen, Antispasmodics) to embark on her journey to treatment.

Lamentably, the effect of treatment was interim and in due course she caved into the unresponsiveness of medical therapy. After a long haul of workup and discussion, staged globus pallidus interna lesioning was planned. A total of four lesioning was instituted on either side but the consequent was only brief and short lived. Despite the surgical venture, the narrative was agonizingly interchangeable as with the precursory oral medications. To add to this letdown, the severity of the stiffness crumbled so much that she needed to be at the mercy of someone else even to commit herself to feeding and ambulating. In an attempt to come to grips with her infirmity, an intrathecal baclofen testing was proposed as a surrogate.

Procedure

There was a dramatic improvement in her symptoms. It was a notion of triumph to see a patient revamping from a state of lying flat on one's back to being self-ambulatory as an unrestrained individual. Unluckily, we were confronted with subcutaneous collection within the abdominal pocket where the system was allocated that required repeated aspiration on many occasions. Fortunately, with broad spectrum antibiotic coverage, when all was said and done the crisis was hell bent.

She is now in a much better shape and her family is as rejoicing as her. Heretofore, we have refilled the baclofen thrice, once in 80 days.

1. A thorough neuropsychological evaluation was executed.
2. A meticulous general and physical assessment was instituted to ensure the fitness for the surgery and exclude pre-existing infections.
3. Test dose of baclofen was given starting off with 100 mcg and increasing up to a dose of 150 mcg over 3 days with an admissible improvement in her neurological status.
4. Intraoperatively, spinal catheter was inserted up to D9 level under C-arm guidance,
5. With assurance of CSF outflow, spinal catheter was anchored in the fascia using fixation wing.
6. 12 ml of Baclofen was diluted in 28 ml of normal saline, making a total of 40 ml (Rate: 0.5 ml/day = 75 mcg/day)
7. 40 ml of that preparation was injected into the pump.
8. Finally, the spinal catheter that was inserted intrathecally was connected to the pump.
9. The whole system was placed in the abdominal pocket and anchored and wound closure was done.

CSF, cerebrospinal fluid.

Discussion

Tardive dyskinesia encompasses a symptom nexus of dystonia in conjunction with stereotypy, akathisia, chorea, myoclonus, and tremor[14]. Several mechanisms have been postulated relating the development of tardive dystonia to the administration of neuroleptic drugs[15–17]. The culpability is believed to be caused by striatal dopamine receptor super-sensitivity persuaded by the dopamine-antagonizing action of antipsychotic drugs. The foremost rationale to treat tardive dyskinesia, on that account, aim to maneuver these dopaminergic pathways in discrete ways. Owing to the postulated reciprocal alliance between striatal dopaminergic pathways and cholinergic pathways, auxiliary therapies have been recommended that exert influence on cholinergic pathways as well[18,19]. Likewise, involvement of γ aminobutyric acid GABAergic pathways has been conferred about[20], interestingly as a feature, possibly typical of tardive dystonia as compared with nontardive dystonia[16]. This has underpinned the foundation to endorse drugs that amend GABAergic transmission, such as the GABAB agonist, the prototype being baclofen[15].

Contingent upon with the patient manifesting signs of generalized dystonia, notably with onset at a young age, and is being considered for treatment with intrathecal baclofen, a preliminary obligation should be acknowledged by the practitioner to rule out primary generalized dystonia[21]. The proceeding may involve consulting a neurologist with movement disorders expertise and/ or dispatching specialized genetic testing for DYT1. If the patient suffers from an early-onset primary generalized dystonia, then treatment with deep brain stimulation of the globus pallidus interna is the approved and preferred treatment approach. Per contra, the common indications for intrathecal baclofen includes spasticity, quadriplegia in cerebral palsy, spasticity, and paraplegia of spinal origin, hereditary spastic paraparesis, and secondary generalized dystonia[8,19].

Besides, there are a handful of contraindications to intrathecal baclofen therapy. These include an allergic reaction to baclofen, poor health precluding general anesthesia, and an

inability to attend the necessary follow-up appointments for refills. Even though the absolute contraindications to baclofen therapy are relatively disparate, there are a myriad of circumstances that a practitioner must contemplate when deciding whether or not intrathecal baclofen is the out-and-out riposte to the patient's ailment. For instance, it is indispensable in the practitioner's part to gage the psychosocial situation and ascertain whether or not the patient can attend follow-up appointments for programming and refills under their own power or if their situation entail a need of significant logistical support[14]. If the patient is incapacitated to the degree that their situation compel assistance with transport and care, one must authorize that they have that support from the nearest kin, before embarking on this blueprint.

Many co-morbid state of affairs that may increase the risk of complications should be ameliorated by foregoing the installment of the apparatus. An underweight patient should have his nutritional status enhanced while an overweight patient should have his diet chart calibrated. Infective pathologies like a pre-existing pressure sore or urinary tract infection should be optimized beforehand to avoid getting drenched into the hazards of septic convolution.

The intrathecal delivery holds an upper hand compared to the other alternatives taking note that it permits a more potent antispasticity effect with decreased side effects at a reduced dosage. Surveying with more than 1000 patients world-wide, treated with continuous intrathecal baclofen has inferred it to be a safe and efficacious plan of action for this condition[22]. In this regard, intrathecal instillation of baclofen given as a bolus or continuously by means of an implantable pump apparatus has long been used comprehensively as a treatment of severe and otherwise intractable spasticity[23,24].

Nonetheless, this approach is not unescorted with complications. The most frequent complications noticed during a test dose were nausea, vomiting, sedation, hypotension, urine retention, seizure, weakness, and headache.

A few complications that can be encountered during CIBI implantation are cerebrospinal fluid (CSF) collection, CSF leak, constipation, headache, infection, hypotension, hematoma, hydrocephalus, worsened gait, urine retention, flipped pump, and catheter revision, with the commonest drawback being a CSF leak.

The frequent reason for pump replacement was infection or arachnoiditis[17,25]. Patient's request, hypermobility with effusion, CSF leakage, and dehiscence were also reported as the causes compelling pump revision. Common causes for catheter replacements are associated with hardware malfunction, such as kinks or catheter migration or occlusion and due to end of battery life[25,26].

Considering the frustration and disappointment despite multiple sessions of lesioning in our patient, installing an intrathecal baclofen pump was the only desperate remedy. Prior to the surgery, the aftermath was ambiguous. Yet the sequel was a jaw-dropper with the patient experiencing a marked reduction in her dystonic symptoms without reduction of the muscle strength or any other substantial adverse aftereffect. She is under regular follow-up, and the triumph we experience to see her as an independent individual is second to none.

Conclusion

In the wake of the long haul with the use of the continuous intrathecal baclofen pump for cases with tardive dystonia refractory to conventional therapy, it has been approved as one of the safest and capable procedures. A diligent workup is imperative before deciding to embark onto this approach. Despite having a prosperous aftermath, we should always bear in mind that this strategy in not untouched with a sweep of complications.

Ethical approval

The case study was cleared by the institutional review committee of Annapurna Neurological Institute and Allied Sciences.

Consent

A written and signed consent for publication has been received from patient and her family.

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Author contribution

J.K.: study design, writing original draft, conceptualization, review and editing; B.S.: conceptualization and editing; P.G.: study design, supervision, review, editing; R.S.: supervision, review, editing; S.D.: review, editing; B.P.: supervision.

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The author reports no declaration of interest.

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