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Foix-Chavany-Marie-Syndrome – Neurological, Neuropsychological, CT, MRI, and SPECT Findings in a Case Progressive for more than 10 Years*

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Summary. In a 66-year-old woman signs and symptoms of bilateral opercular syndrome (Foix-Chavany-Marie-syndrome) developed progressively over a period of more than 10 years. Facio-linguo-velo-pharyngeo-masticatory diplegia with automatic-voluntary dissociation was accompanied by motor aphasia and oral apraxia leading to a state of almost complete anarthria. Although it initially resembled the anterior biopercular syndrome there are also features indicating involvement of the posterior opercula. Although the aetiology remains obscure without pathological data, a bilateral focal brain atrophy is assumed. This is probably the first case documented by MRI and SPECT.

Key words: Opercular syndrome – Pseudobulbar palsy – Neuropsychological findings – Brain imaging

Introduction

In 1926, Foix, Chavany and Marie (Foix and Chavany 1926; Foix et al. 1926) described a cortical variety of pseudobulbar palsy termed facio-linguo-velo-pharyngo-masticatory diplegia, ascribing its origin to lesions in the territory of the precentral branch of the middle cerebral artery bilaterally. A key diagnostic sign was automatic-voluntary dissociation with selective impairment of volitional movements but without affecting automatic and reflex motor activity.

Several varieties of Foix-Chavany-Marie (FCM) syndrome have been described: accompanied by complete speechlessness (speech suppression) without aphasia (Alajouanine and Thurel 1933; Arnould et al. 1967; Cappa et al. 1987; Colombo et al. 1983; Mariani et al. 1980; Villa and Caltagirone 1984), by aphasia and/or oral apraxia (Colombo et al. 1983; Ferrari et al. 1979; Levine and Mohr 1979; Mariani et al. 1980; Sandyk and Brennan 1983; Sandyk 1983), and by pure word deafness (dalla Pria et al. 1979). Depending on the relative involvement of the anterior (Biller et al. 1981) or the posterior operculum, two varieties of the syndrome are distinguished. The term FCM syndrome is often reserved for the anterior variety (Arnould et al. 1967). About 30 cases had been reported in the literature up to 1983 (Mariani et al. 1980; Sandyk 1983; Bruyn and Gathier 1969); these are almost exclusively considered to be of vascular origin (Arnould 1967 et al.).

CT shows more or less symmetrical bilateral infarctions (Mariani et al. 1980; Colombo et al. 1983; Villa et al. 1984; Levine and Mohr 1979; Ferrari et al. 1979; dalla Pria et al. 1979) or tissue loss with marked brain atrophy (Sandyk 1983; Sandyk and Brennan 1983). This paper reports the case of a patient presenting with a primary bilateral opercular syndrome that has been slowly progressing for over 10 years.

Case Report

In a right-handed woman with 8 years' school education, mild dysarthria of insidious onset was noted when aged about 50 years, combined with some difficulty in writing. At the age of 55, dysarthria of the bulbar type was marked, articulation laborious and variable, and syllables were omitted or blurred. The patient herself noted mild word finding difficulty. Mentation was rather slow, but memory was undisturbed. EEG, brain scintigraphy and CSF were normal. The disorder was initially thought to be of psychogenic origin.

At age 58 dysarthria was so pronounced that the patient preferred to write, when she occasionally confused letters or

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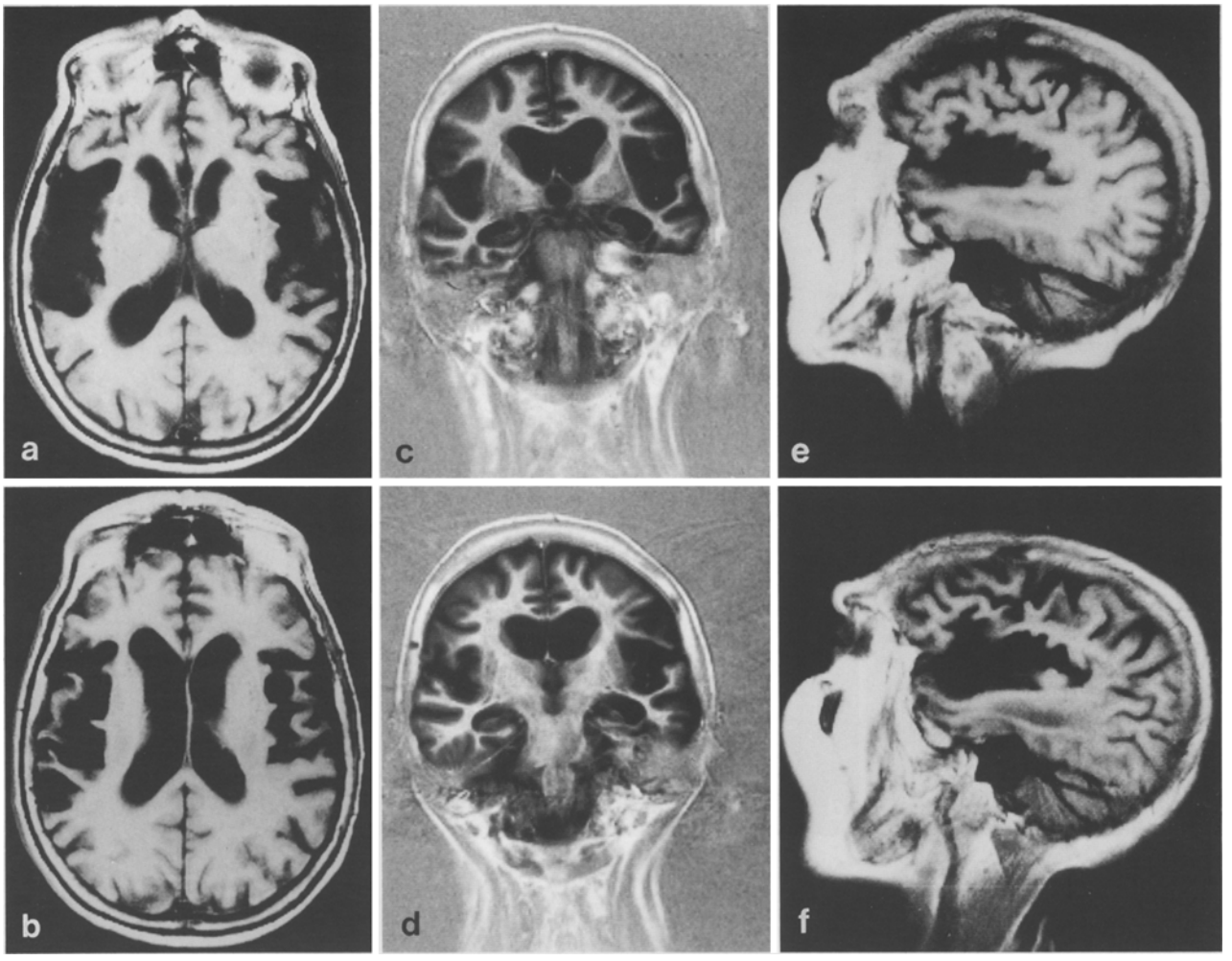

Fig. 1a–f. MRI scans at the age of 66 years show tissue loss of the opercular region more pronounced on the left side, also visible on the coronal and sagittal sections

omitted words. She complained of a feeling of constriction in her throat, which was relieved when she ate. Singing became almost impossible because of dyspnoea. She felt that she was drooling with the right angle of her mouth. No neurological abnormality other than dysarthria was noted at that time. Mentally she was fully oriented and cooperative. Personality testing showed abnormally high scores on "hypochondria", "depression", and "hysteria" (Hathaway and McKinley 1951). CT scan was normal.

At the age of 61 she presented with chronic tracheobronchitis and coughing. On neurological examination the soft palate was lifted incompletely, her speech was still markedly dysarthric and alternating movements of the right hand were slightly disturbed. Dysgraphia in spontaneous writing had worsened through orthographic mistakes and agrammatical tendencies; writing to dictation and word synthesis by letters was, however, fairly well preserved. Auditory and written comprehension were good, and Token Test was normal. Spontaneous speech was marred by phonemic paraphasias which were thought to be partially related to oral apraxia. The patient was unable to imitate oro-bucco-facio-linguo-pharyngeal movements or to perform them according to command; parapraxias occurred. Articulation was not improved by tactile, visual or auditory cues. Performance of oral repetition, reading aloud and naming was similar to spontaneous speech. The neuropsychological diagnosis was motor aphasia, accompanied by oral apraxia. On otorhinolaryngologic examination larynx function was normal. CT scan now showed a widened left insular cistern.

At the age of 66 she was again hospitalized having suffered two attacks of severe asphyxia because of her inability to expectorate mucus or swallow appropriately. She complained about difficulty in swallowing, increased saliva production and desire to urinate. She had lost 20 kg during the past 2 years. She could not close her eyes on command, but could lift her eyebrows slightly; both angles of her mouth were depressed, but she could raise them when she laughed. She yawned normally. The mouth was slightly opened and the mandible could be moved only about 2 cm actively. Interestingly she was able to open her mouth when she was asked to stick out her tongue. There was drooling, the tongue was almost immobile but without atrophy, and the masseteric reflex was brisk. The corneal reflex was unremarkable and gag reflex was absent. The glabella reflex, snout reflex and palmomental reflex were brisk. The velum was immobile on inspection. The mobility of the vocal cords was reduced, the glottis was wide open, and there was excessive mucus accumulation in the trachea. An irregular triggering of the act of swallowing was noted. On manometry pharyngeal transport was irregular and the tone was reduced.

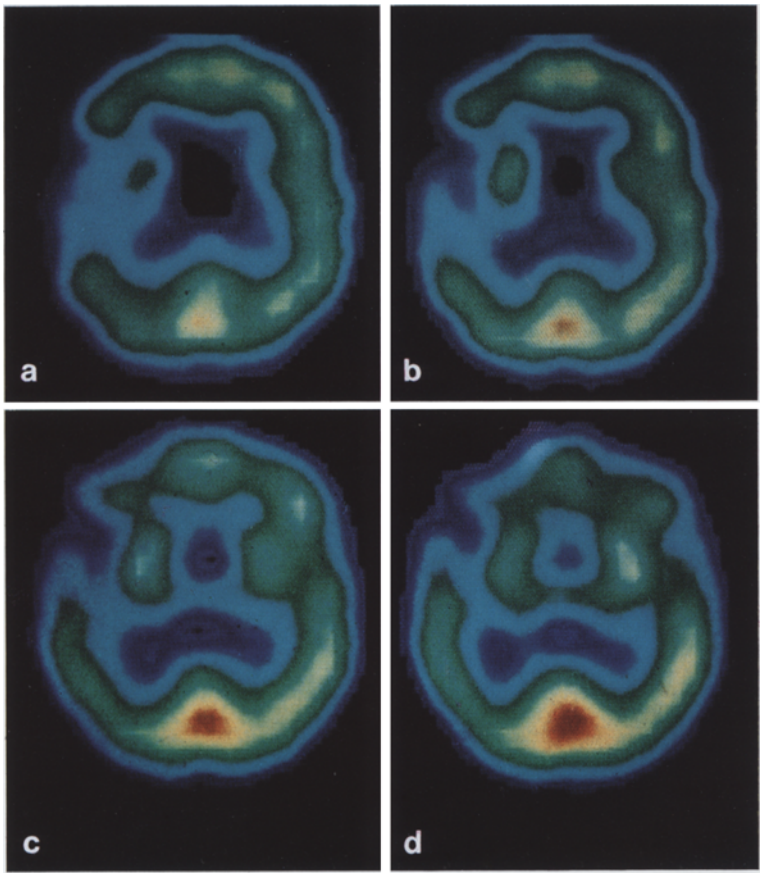

Fig. 2a–d. ^{99}Tc-HMPAO single photon emission tomography showed markedly reduced perfusion of exactly the left and (to a lesser extent) of the right opercular region

During her stay on the ward she ate almost normally. She was completely speechless except for moaning sounds and communicated exclusively with the aid of a typewriter. An audiogram yielded mild symmetrical bilateral high frequency loss. No abnormalities of the extremities were noted. Mentally she was fully awake and oriented; she had no difficulty in finding her way on the ward. EEG showed some bilateral flat theta waves in the precentral regions. CT revealed widespread atrophy of the brain, more pronounced on the left side and in the sylvian regions without circumscribed infarcts or signs of subcortical arteriosclerotic encephalopathy. Perisylvian and insular tissue reduction was visualized much better on MRI, which did not show significant subcortical changes indicative of vascular disease.

^{99}Tc-HM PAO-SPECT showed marked reduction of activity in the left opercular region, much more than on the right side.

Following another aspiration she was fed intravenously and consequently supplied with a percutaneous endoscopic gastrostomy, which led to considerable weight gain.

Neuropsychological Assessment. Detailed testing was carried out between November 1986 and February 1987. Several functions were difficult to assess because of a complete inability to speak.

The patient was cooperative with little sign of fatigue. When indicating a yes or no answer she used hand signs instead of nodding or shaking her head. On some of the tasks she was slow and undecisive. She was able to indicate the day of the week on a scale and was apparently oriented with regard to place. The results of neuropsychological testing are summarized in Table 1. Short-term memory was tested by a self-developed continuous figure recognition test which requires no overt verbal skills (Lang 1989). Spontaneous speech, reading aloud and oral repetition could not be tested because of almost complete anarthria.

There were extensive spontaneously handwritten reports from 1975, 1978, and 1982; the report of 1986 was typed. We noted an increasing number of omissions and other literal (additions, substitutions, transpositions), syllabic and verbal paragraphias.

The syntax became more and more agrammatical; finally the handwriting was barely legible and the meaning had to be guessed.

At the last examination she was able to copy letters but made mistakes on a letter level when writing to dictation. She wrote graphically similar letters or added, substituted, omitted or distorted letters within words. She was not able to write her name legibly or correctly. Also she was unable to assemble printed letters correctly to make a word, getting half of the letters right at the most. On a sentence assembly task she almost inevitably omitted one word or syllable. She was not helped by pointing to letters of the alphabet instead of writing. She could not indicate how many syllables or letters a word should have.

Discussion

FCM syndrome has usually been reported as a result of secondary contralateral damage with a primary unilateral opercular lesion (Mariani et al. 1980; Cappa

Table 1. Summary of neuropsychological testing

Function	Test	Result (raw, sum score, if not indicated otherwise)
Memory	CFR-pictures	
	geometric	18/22
	figures	3/22[a]
Intelligence	MWT-B	70 (IQ)[a]
	RSPM	90 (IQ)
Flexibility and categorization	WCST-categories	2/6[a]
	errors	27/64[a]
Auditive discrimination	Tonal memory	8/14[a]
	Noise identification	6/10[a]
Arithmetics	Addition/subtraction	0/10[a]
Construction and topographic orientation	Drawing	Without details but otherwise fairly normal
	Clock reading	6/10[a]
	Koh block test	1/12[a]
Language	AAT spontaneous speech	Not testable (anarthric)
	Token Test	33/50[a] (marked deficit)
	Repetition	Not testable (anarthric)
	Writing and reading	4/90[a] (severe deficit)
	Naming	78/120[a] (marked deficit, multiple choice testing)
	Comprehension	63/120[a] (marked deficit)
Praxis	Oral	0/10[a]
	Ideomotor	4/10[a]
	Ideational	5/10[a]

CFR = Continuous figure recognition; MWT-B = multiple choice vocabulary test; RSPM = Raven's standard progressive matrices; WCST = Wisconsin card sorting test; AAT = Aachen Aphasia Test

[a] Subnormal as compared with a healthy age-matched control group ($n = 6$ to $n = 24$) or with standard norms

et al. 1987; Bruyn and Gathier 1969; Crumley 1979; Schott et al. 1961). In our case, however, there was evidence of a primary bilateral opercular pathology progressive for more than 10 years, so that the question arises whether there might be an underlying primary focal brain atrophy if one is not to assume a steadily progressive vascular process confined to the opercular cortical branches of the middle cerebral arteries. There was no clear evidence of an insult-like event in the patient's history and vascular risk factors were lacking. The only other case we found in the literature which was of insidious onset at the age of about 50 years and progressed further is that of Arnould et al. (1967). The steady progression in our patient was demonstrated most impressively by dysarthria and dysgraphia. However, it cannot be denied that hypoxic damage through recurrent asphyxia reminiscent of what has been described previously as periodic laryngeal dyspnoea (Cambier et al. 1983) may have contributed to her present status. Neurological findings, i.e. mainly dysarthria, difficulty in deglutition and facial palsy were indicative of the bilateral involvement deemed characteristic of a fully fledged FCM syndrome, somewhat more pronounced on the left side. Furthermore, voluntary/automatic dissociation (Bruyn and Gathier 1969) was an important hallmark. The patient's face was motionless, eyes and mouth were open, there was drooling, and hardly any movement on command, while on emotional reactions the angles of the patient's mouth were lifted and stretched out substantially together with a slight narrowing of the palpebral fissure and some unformed phonation. This may be explained by intact limbic system connections. When asked to demonstrate eating she was unable to do so, but when she actually ate there was scarcely a noticeable difference from a normal eater except for a disturbance in initiating the act of swallowing. Of the three stages of swallowing – voluntary, pharyngeal and oesophageal – the first was possible only when actually food was present, and the transition to the second stage was disturbed. Remarkably she could not open her mouth on command but did so when asked to stick out her tongue (which she was unable to do). Production of saliva as an autonomic function mediated by the eighth nerve, and sensory fifth nerve functions were unaffected.

While in subcortical pseudobulbar palsy, exaggeration of the masseteric reflex can be demmonstrated as a rule, it was normal in our patient. Gag reflex was weakened. Palmomental reflex could be elicited bilaterally. Despite facio-lingual palsy there was no atrophy, fibrillation or fasciculation as seen in motor neuron disease.

The fact that, on SPECT scan, activity was more reduced on the left than on the right side, fits well into the neurological and neuropsychological findings, which disclosed more left hemispheric (aphasia, apraxia) than right hemispheric deficits. Constructional apraxia may be easily explained by bilateral temporal involvement (dalla Pria et al. 1979), although it more resembled the left hemispheric type. Language comprehension was distinctly disturbed only in the late stage of the disease. Analysis of writing yielded mostly omissions of parts of letters or linguistic units (letters, syllables, words) whose frequency was increasing over time. Since initially more function words than nouns were omitted the written language adopted an agrammatical quality that in combination with initially preserved comprehension and dysarthria suggested motor aphasia as the most probable type of language disturbance. One patient (case 5) of another report (Mariani et al. 1980) also exhibited some "expressive aphasic errors" (mostly literal paragraphias) in her written language and also showed facial apraxia. Since, in the middle of our observation period, oral apraxia was also present in our patient and its localization is ascribed to the frontal and central operculum, first central convolution, and the anterior portion of the insula, there was clear evidence of an anterior opercular syndrome. Persistent nonfluency in Broca's aphasia is associated with involvement of this area plus the underlying white matter (Cappa et al. 1987). While unilateral deficits caused by vascular lesions may clear within a short period of time, bilaterality apparently prevents functional compensation.

Over the years comprehension deficits and ideomotor apraxia emerged, which pointed to an involvement of the posterior operculum, too. Her type of aphasia may now, even acknowledging the complete absence of spoken language, be best classified as global aphasia which is often accompanied by acalculia, constructive deficits, and apraxia. Relative sparing of drawing abilities and absence of severe global memory deficits differentiate the syndrome from temporo-parieto-occipital and medio-temporal-basal pathology as met with Alzheimer's dementia and Korsakoff's syndrome. Whereas neurological signs were a key finding leading to the diagnosis of the syndrome, neuropsychological assessment was of importance in further defining the extent and laterality of the affected brain areas as confirmed by MRI and SPECT scan in particular.

Since focal or labor atrophy is a hallmark of Pick's disease, it may be hypothesized that our case represents a variant of this disorder. In view of the lack of neuropathological data, Alzheimer's disease, focal atrophy due to vascular disease, a non-Pick-non-Alzheimer degenerative condition or even slow virus disease cannot be ruled out.

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